Sounds like a plan!

Steps to improve hearing and communication outcomes for individuals with Down syndrome across the lifespan.

Heather Porter, AuD, PhD, CCC-A
Boys Town National Research Hospital
North Carolina Down Syndrome Conference
April 22, 2023

BOYS TOWN National Research Hospital®

Project INCLUDE

Community Partners

Project INCLUDE

www.boystownhospital.org/ProjectINCLUDE

Scan me

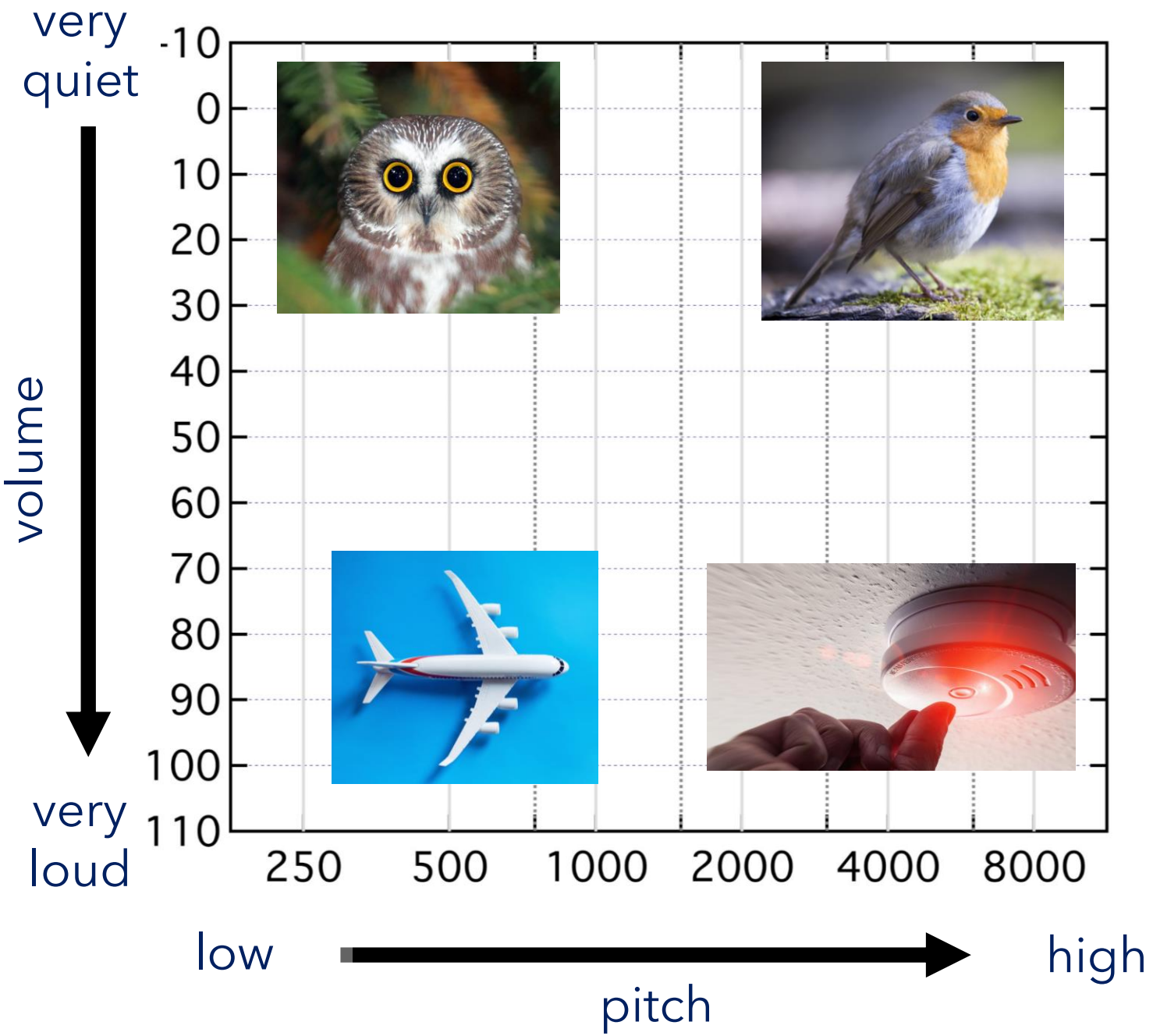
Pick the label that best describes you.

Outline

- Consequences of hearing loss
- Current recommendations for diagnosis, monitoring for hearing loss, and intervention
- Common barriers to good hearing

GOAL
Promote advocacy of evidence-based hearing healthcare services.

Consequences of hearing loss


very quiet
volume
very loud
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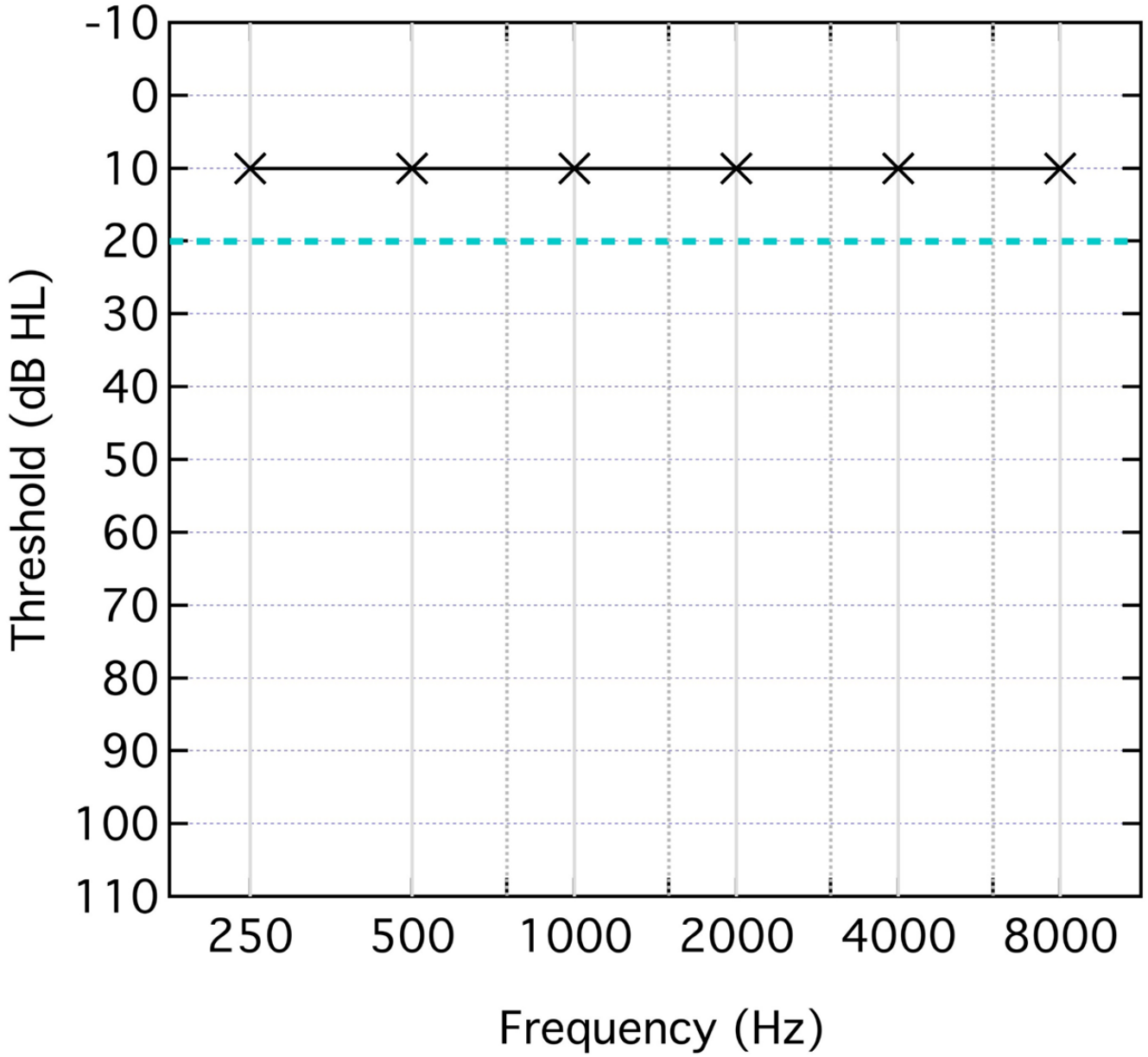


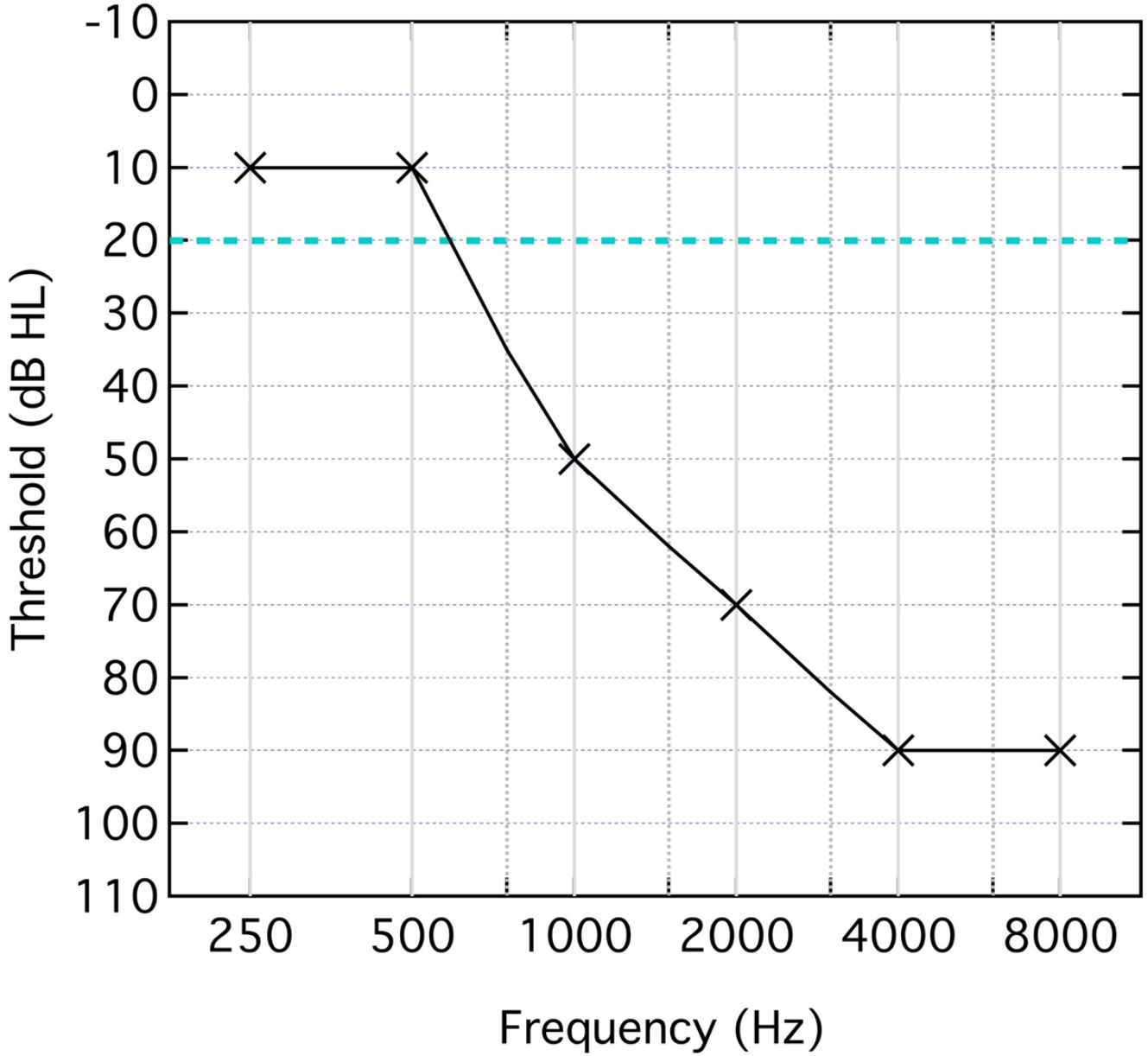
normal

mild

moderate

severe

profound



normal

mild

moderate

severe

profound

Untreated Hearing Loss

- Delayed language development
- Loss of ability to communicate with others
- Social isolation, loneliness, frustration
- Lack of educational progress
- Reduced vocational options

World Health Organization, 2023

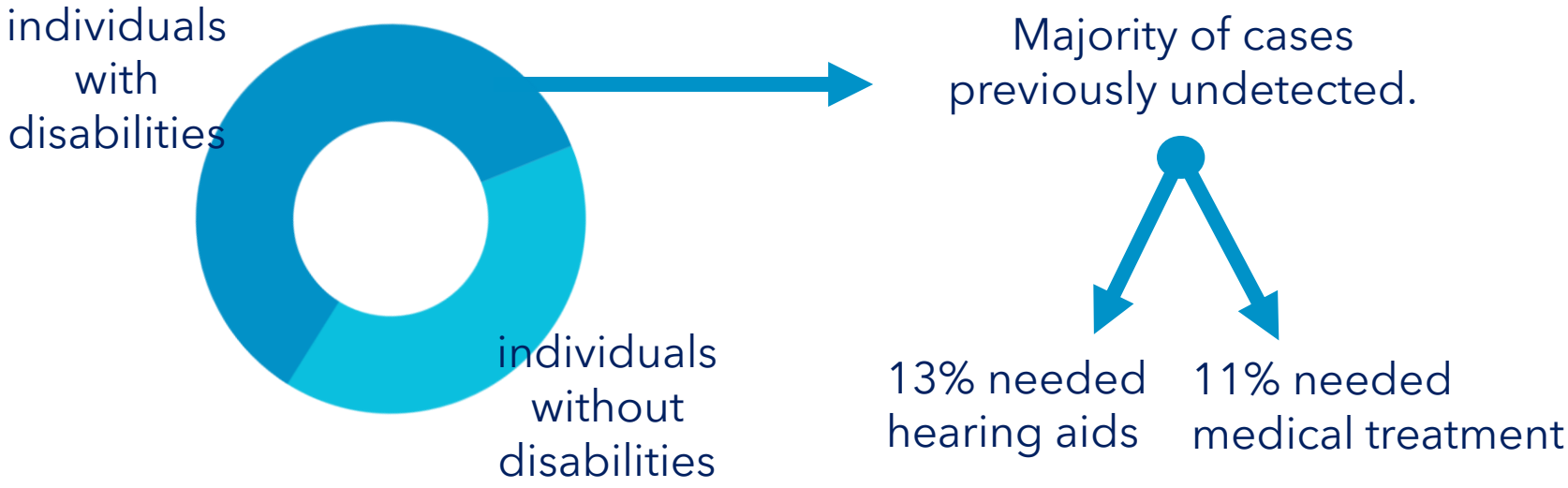
- Intervention benefits children with hearing loss who have additional disabilities and those with hearing loss who have no additional disabilities.

e.g., Cupples et al., 2018; Yoshinaga-Itano et al., 1998

What do think is the worst consequence of untreated hearing loss?

Untreated Hearing Loss: A Special Olympics Report

Hearing Loss Prevalence in the United States



Hearing loss occurs 1.4 times more often in individuals with disabilities than in individuals without disabilities.

Herer, 2012

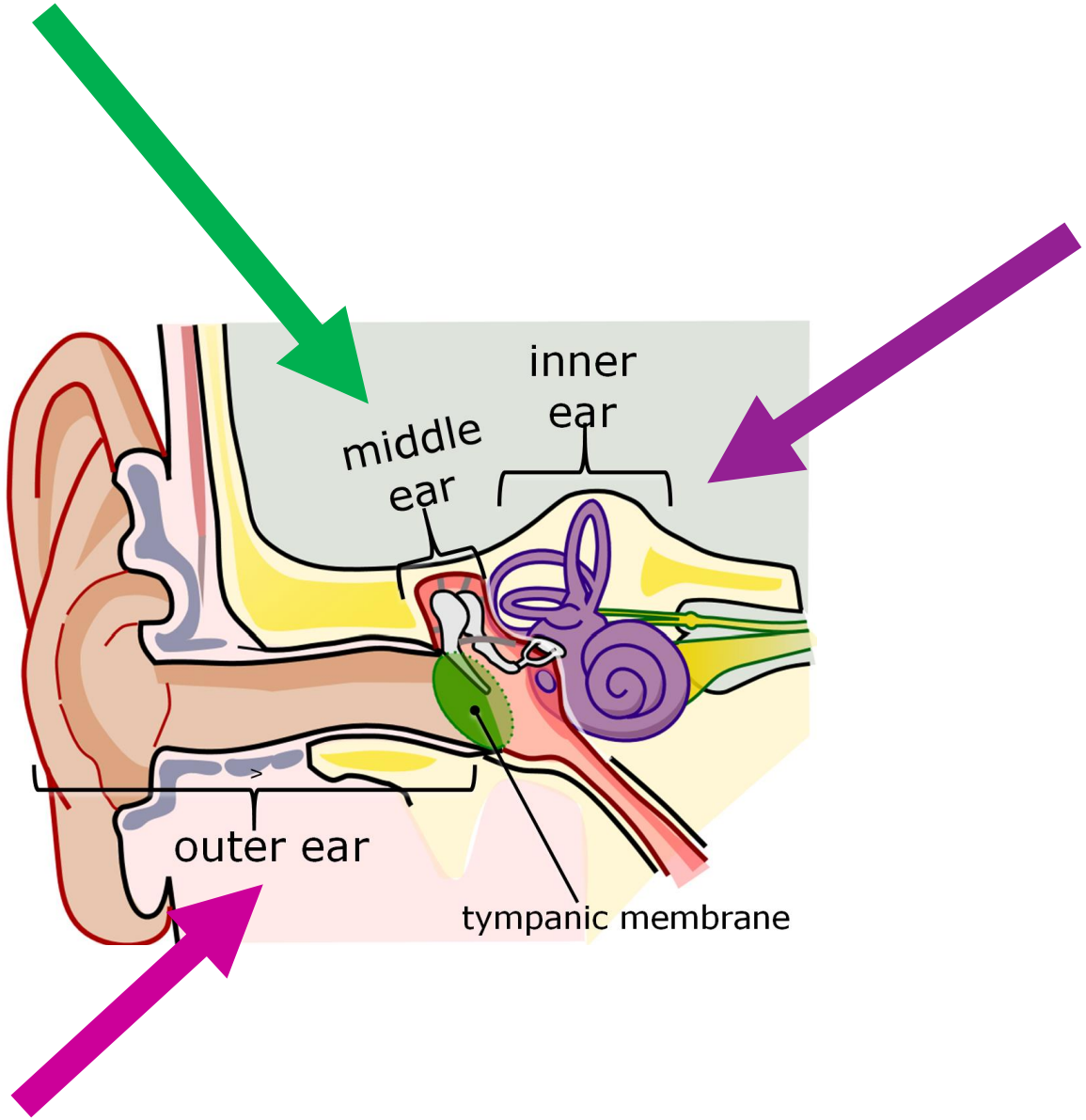
Is all hearing loss the same?

No way!
Ears and hearing are very personalized.

Describing the Unique Characteristics of Ears and Hearing

Describing the Unique Characteristics of Ears and Hearing

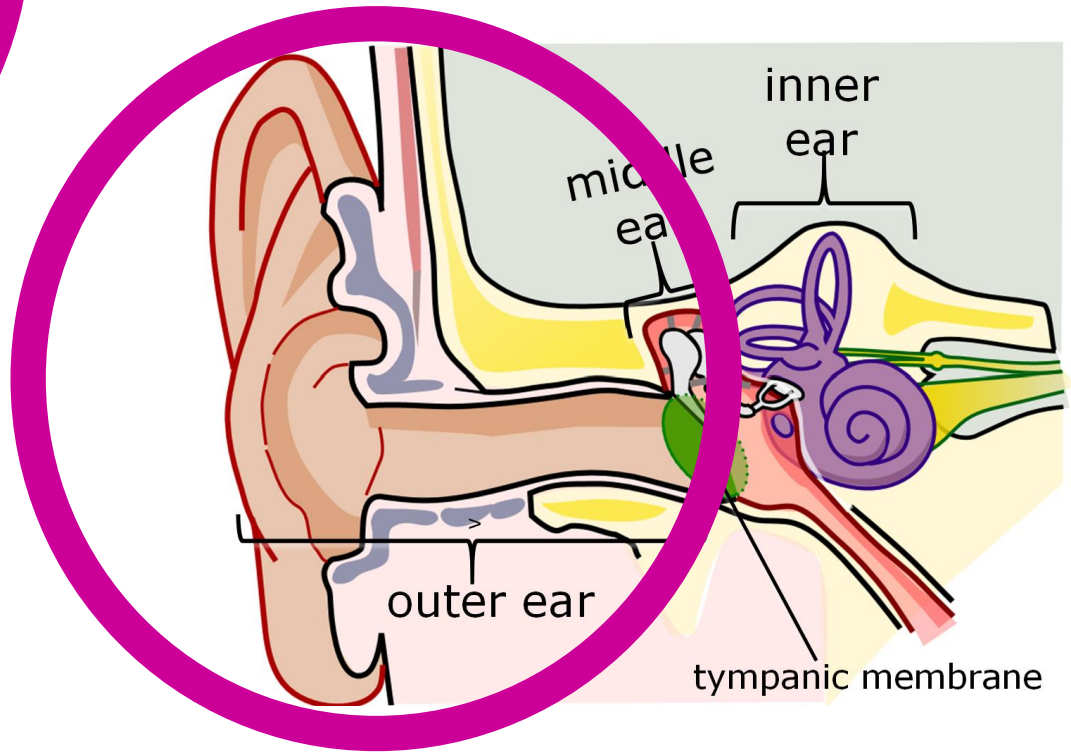
The ear is made up of three sections.



The Outer Ear

Sound is collected by the outer ear and sent to the middle ear by the eardrum.

1.



Outer Ear Tests

Otoscopy

A way to look into the ear and observe the ear canal and ear drum.

Outer Ear Tests

Probe microphone measures

Measures the loudness of sound in the ear.

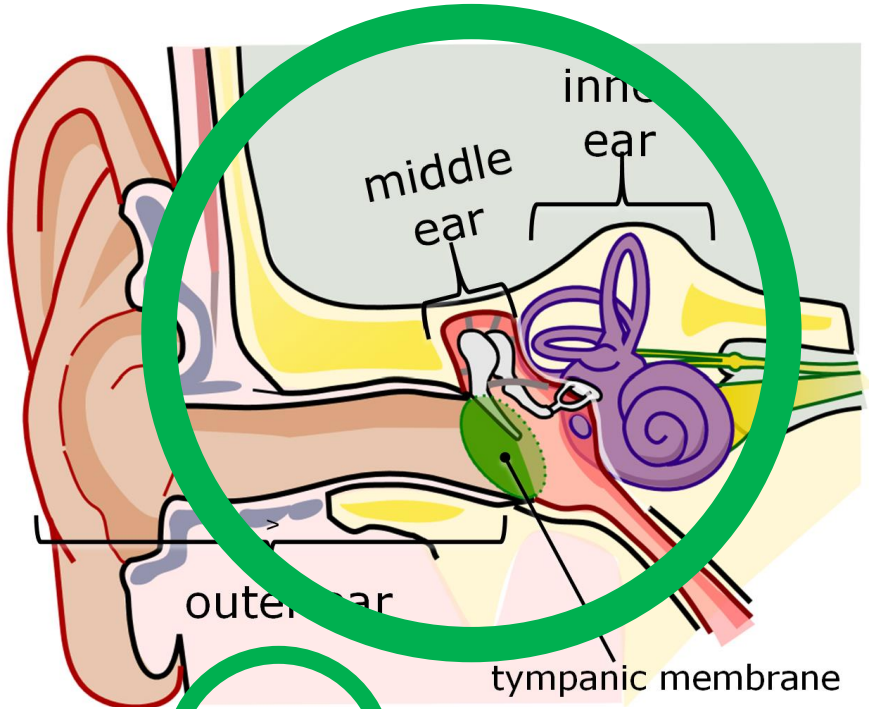
Outer Ear Tests

Probe microphone measures

Measures the loudness of sound in the ear.

The Middle Ear

The middle ear uses the movement of the eardrum and middle ear bones to push vibrations into the inner ear.



2.

Middle Ear Tests

Tympanometry and Wideband Acoustic Immittance

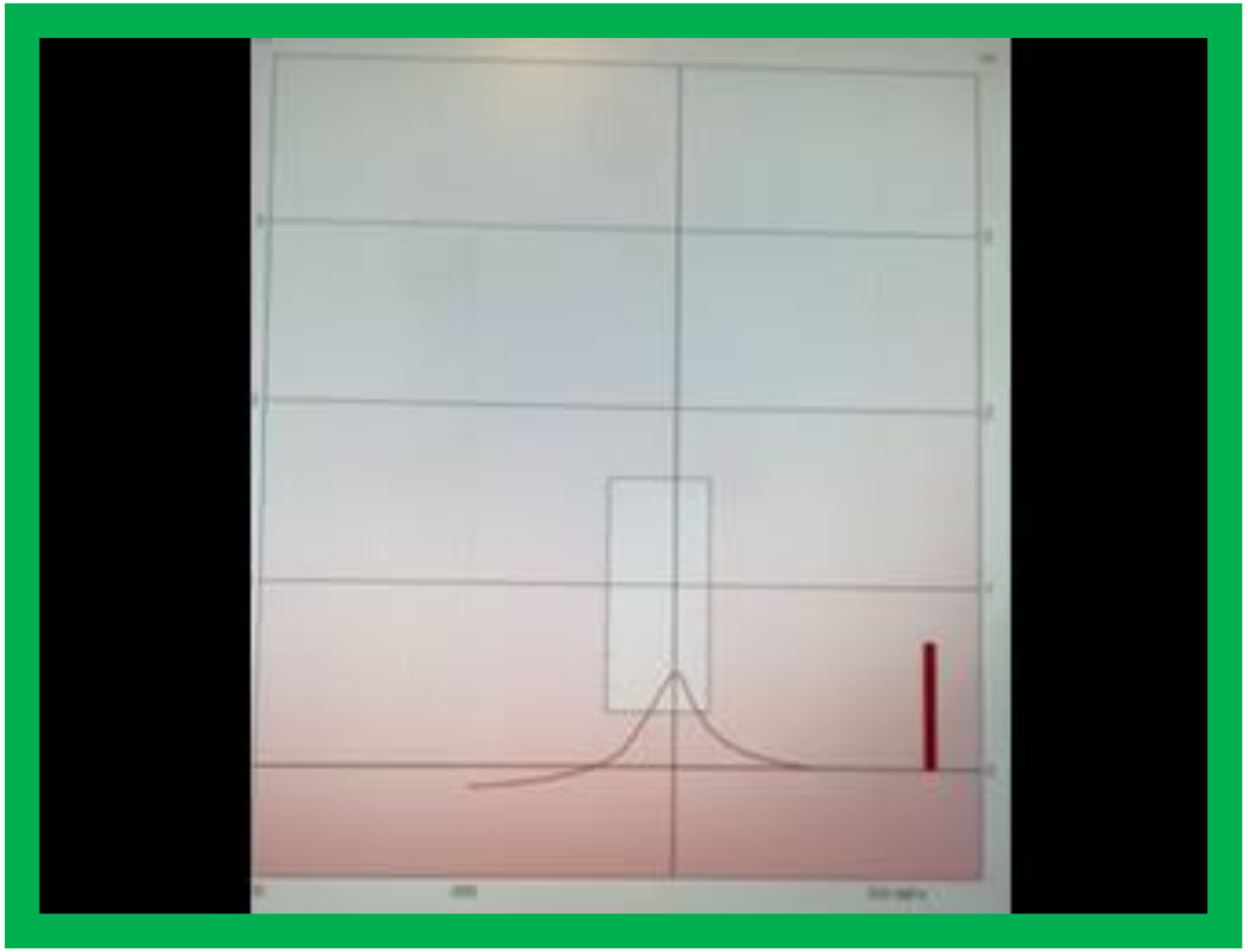
evaluate ear health and can help to determine if there is fluid behind the eardrums

Tympanometry and wideband acoustic immittance

evaluates middle ear health and can help to determine if there is fluid behind the eardrums

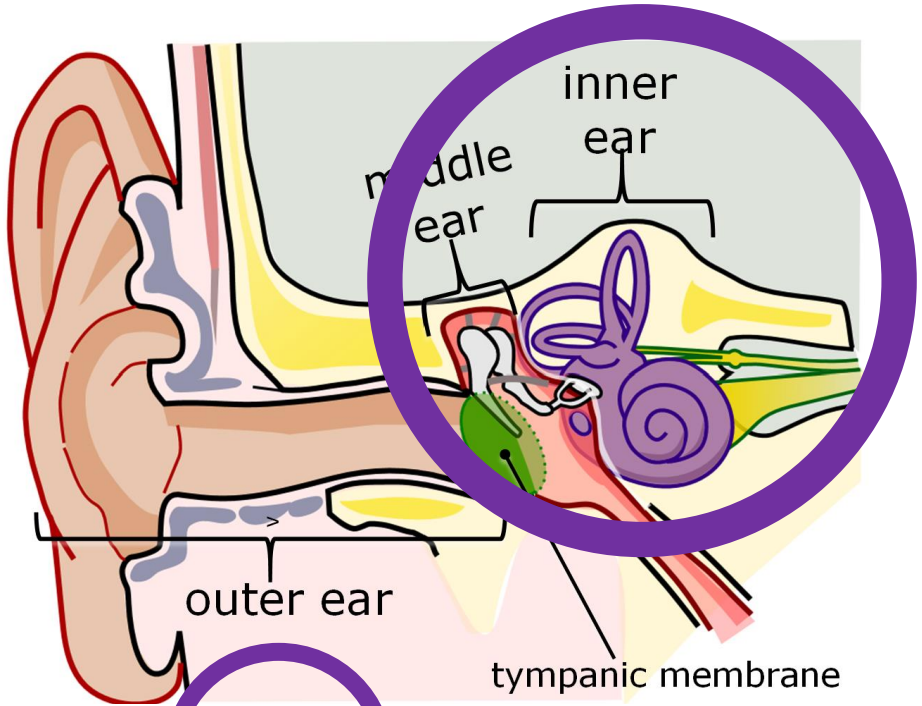
Tympanometry and wideband acoustic immittance

evaluates middle ear health and can help to determine if there is fluid behind the eardrums



The Inner Ear

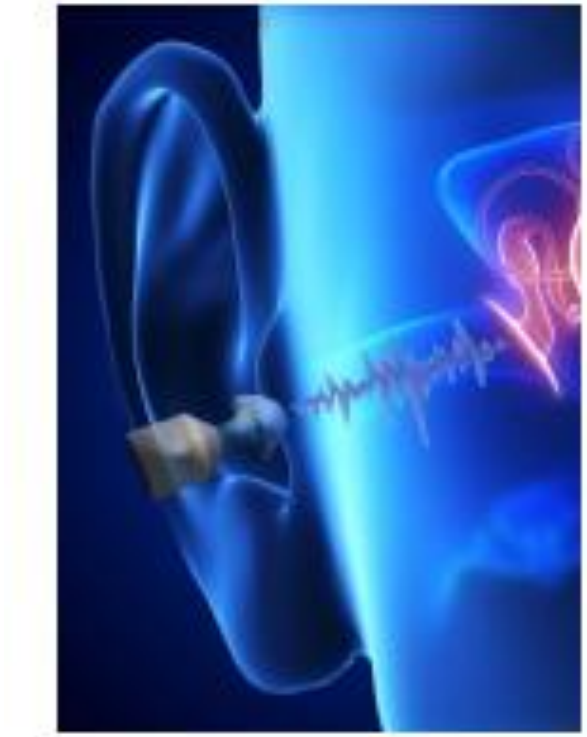
The inner ear codes information and sends neural signals to the brain.



3.

Inner Ear Tests

Otoacoustic Emissions Testing



Sounds that are played into a healthy ear will ‘echo.’ If an ear is unhealthy or has hearing loss, the ‘echo’ will not be present.

Otoacoustic emissions

- Also known as OAE's
- Screen for possibility of hearing loss
- NOT able to measure hearing sensitivity

Neural Tests

Auditory Brainstem Response

This test is performed by observing how the brain responds to sounds.

Auditory brainstem response (ABR)

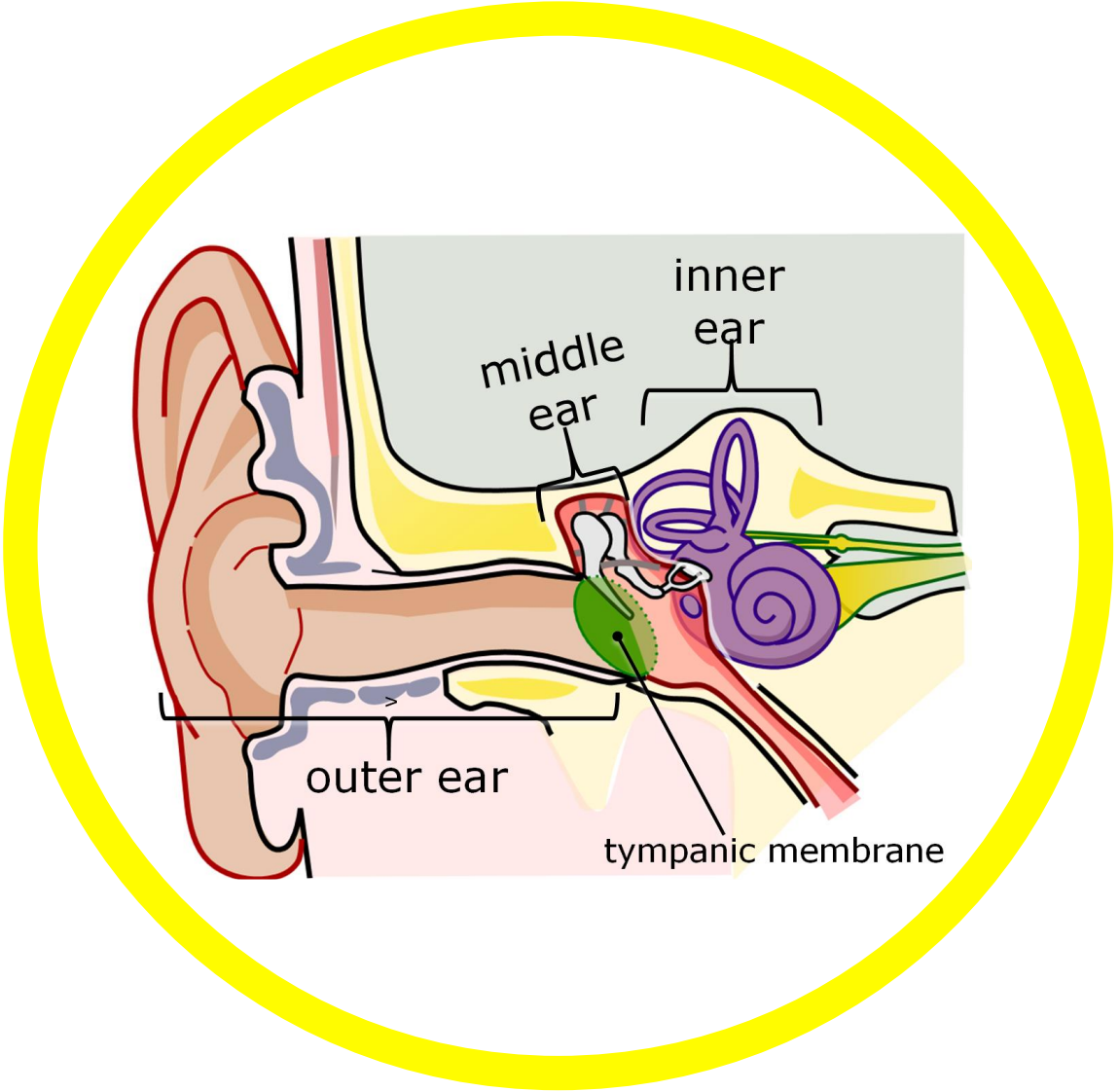
This test is performed by observing how the brain responds to sounds.

Neural Tests

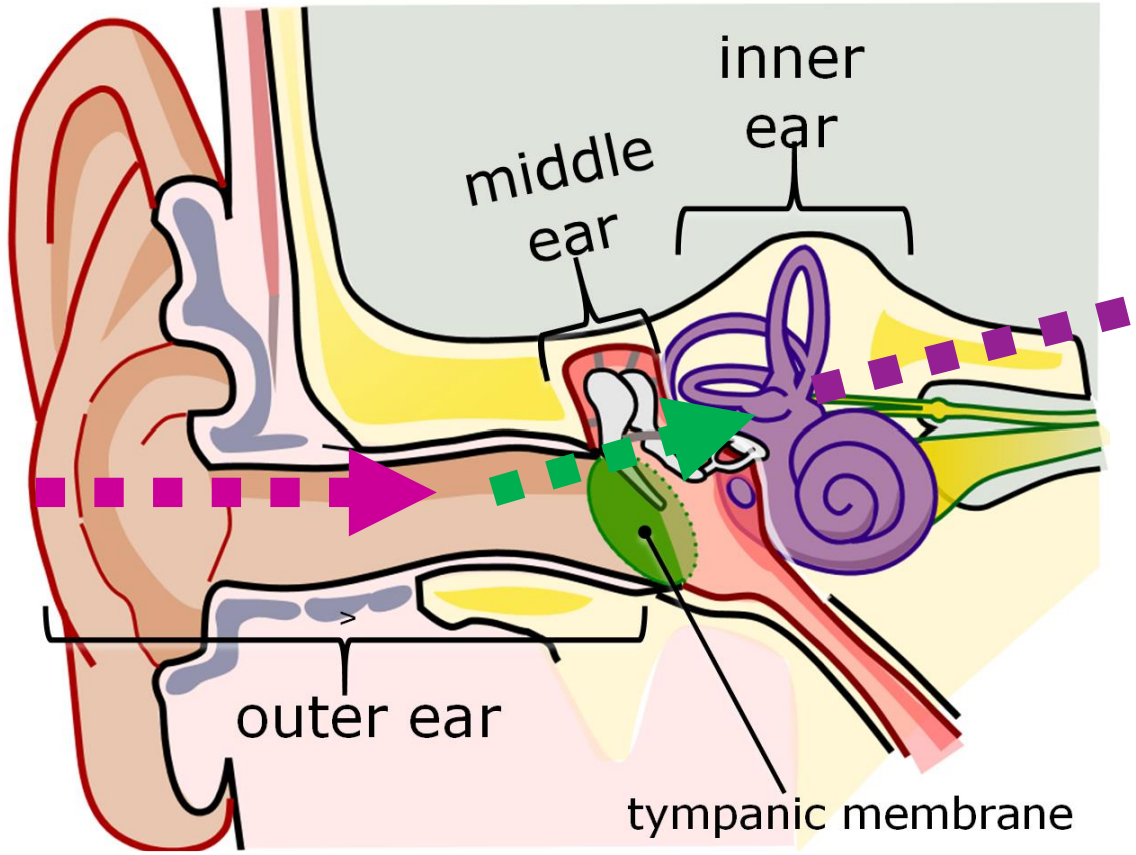
Auditory Brainstem Response

- Also known as ABR testing
- Can be used to screen for possibility of hearing loss
- YES, it can be used to estimate hearing sensitivity

Measuring All Parts of the Auditory System



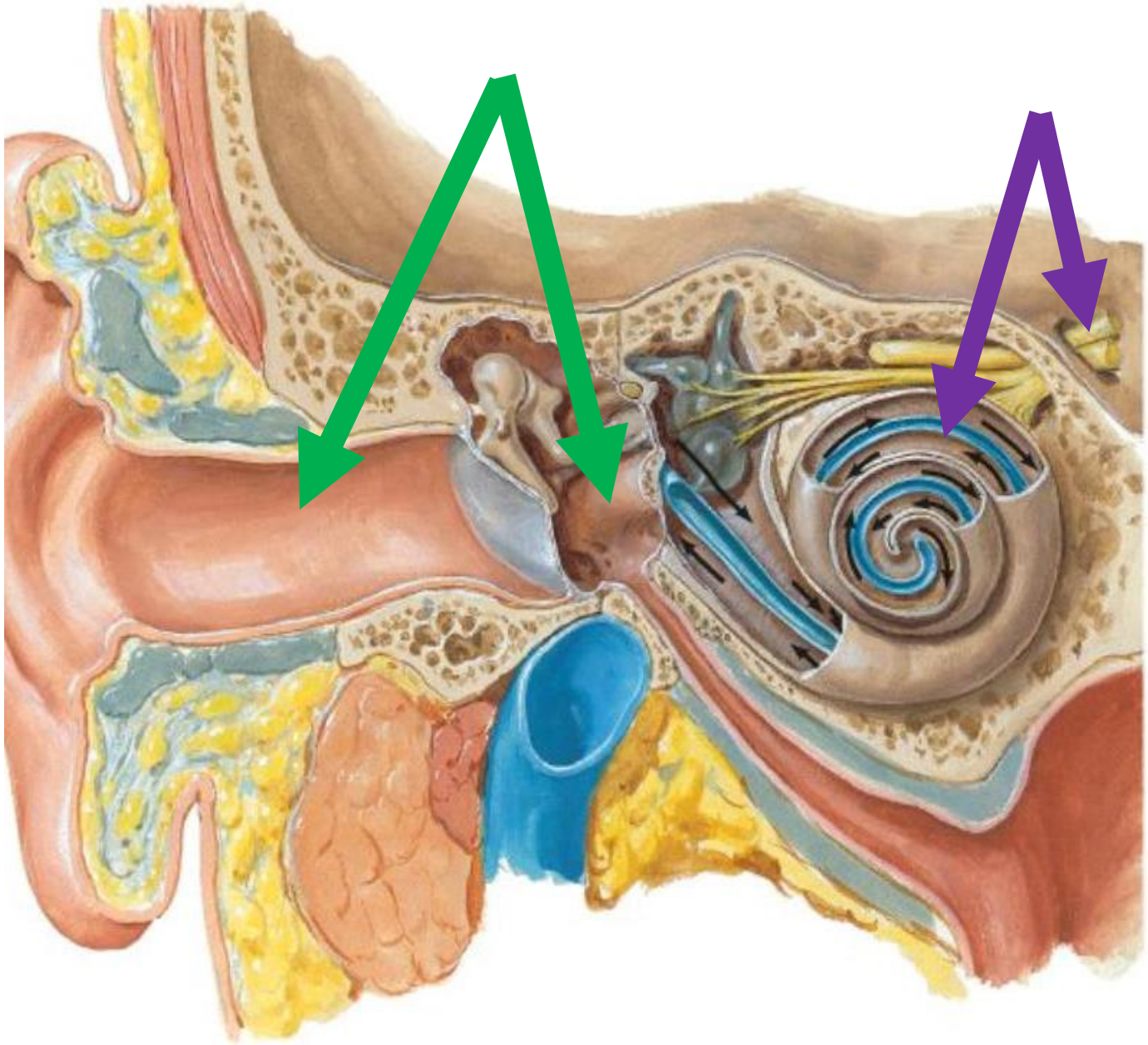
Behavioral Hearing Tests



Behavioral hearing tests

This test requires reliable behavioral responses to sound.

Describing the Unique Characteristics of Ears and Hearing

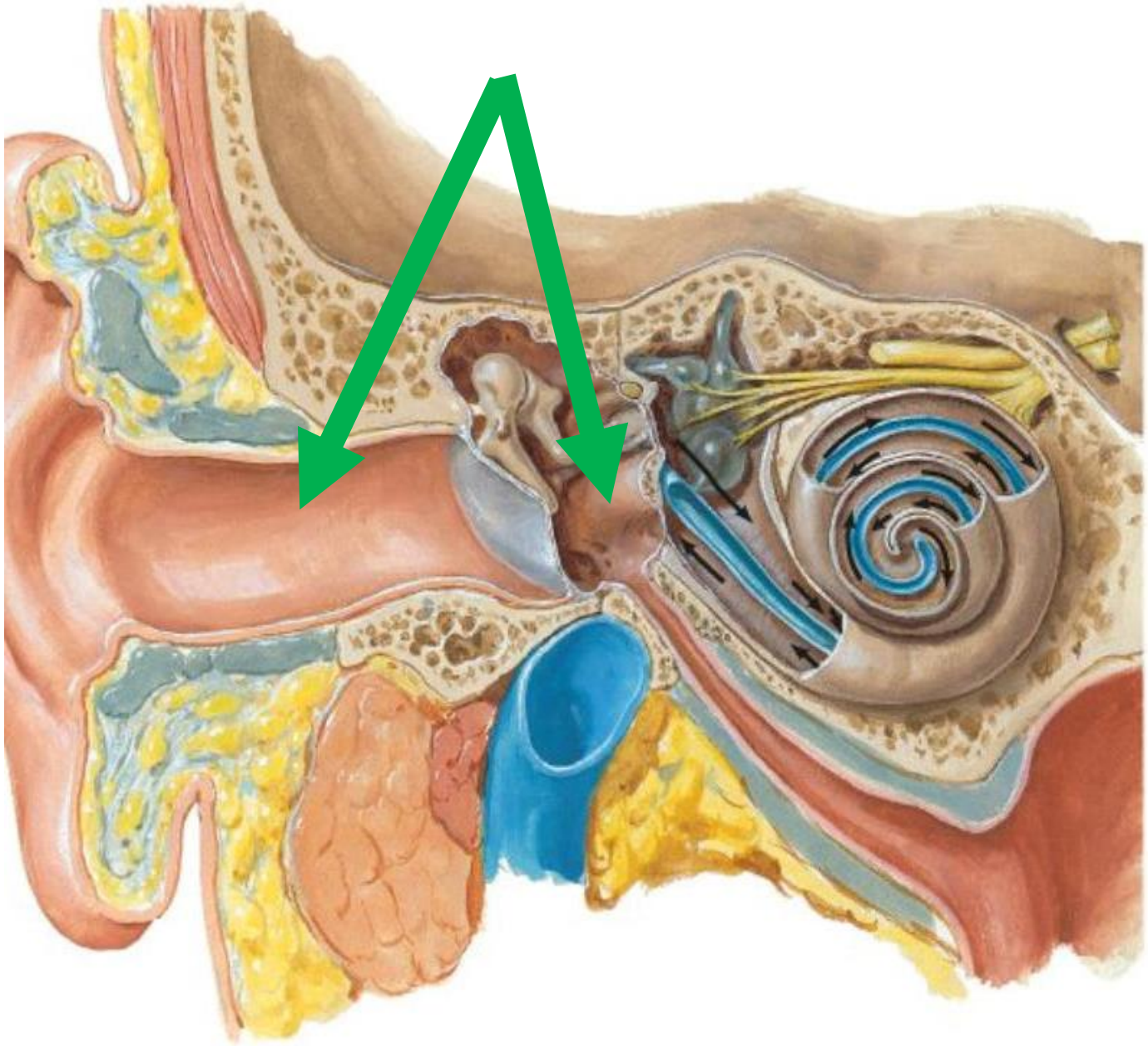


Types of hearing loss

Conductive

Sensorineural

Mixed

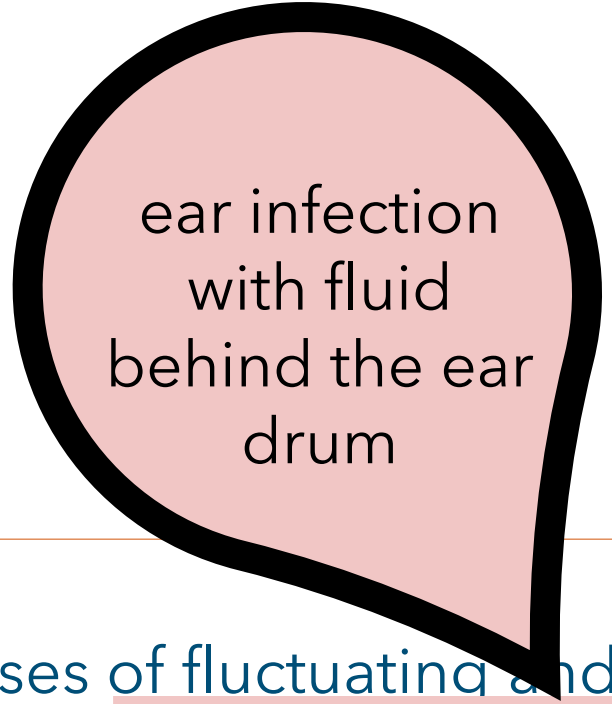


Conductive hearing loss

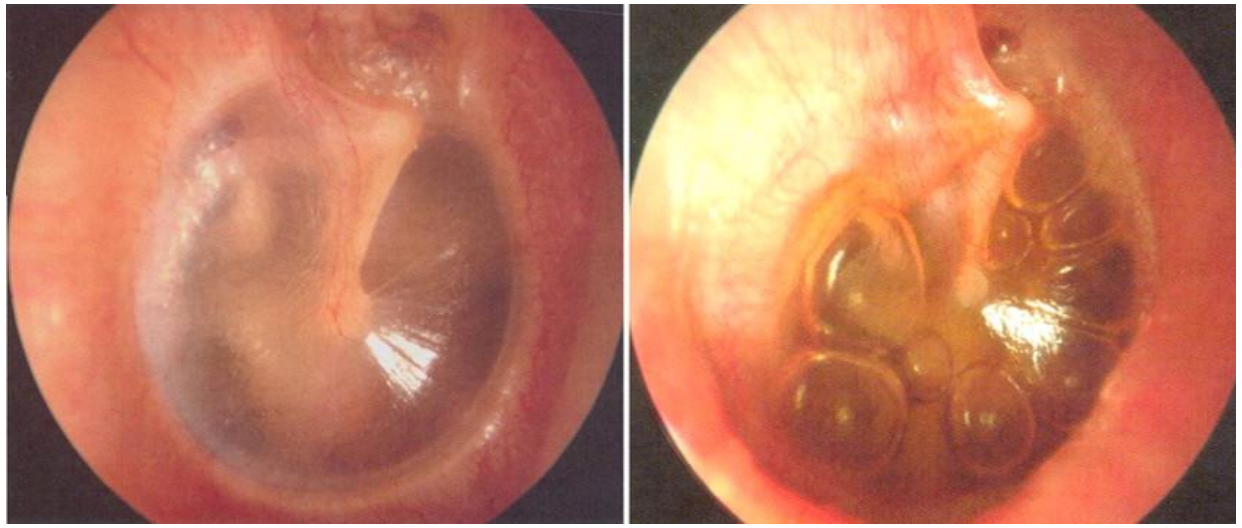
High Rates of Hearing Loss

Occurs in up to 80% of individuals with Down syndrome (Shott, 2006; see Porter & Tharpe, 2010 for review)

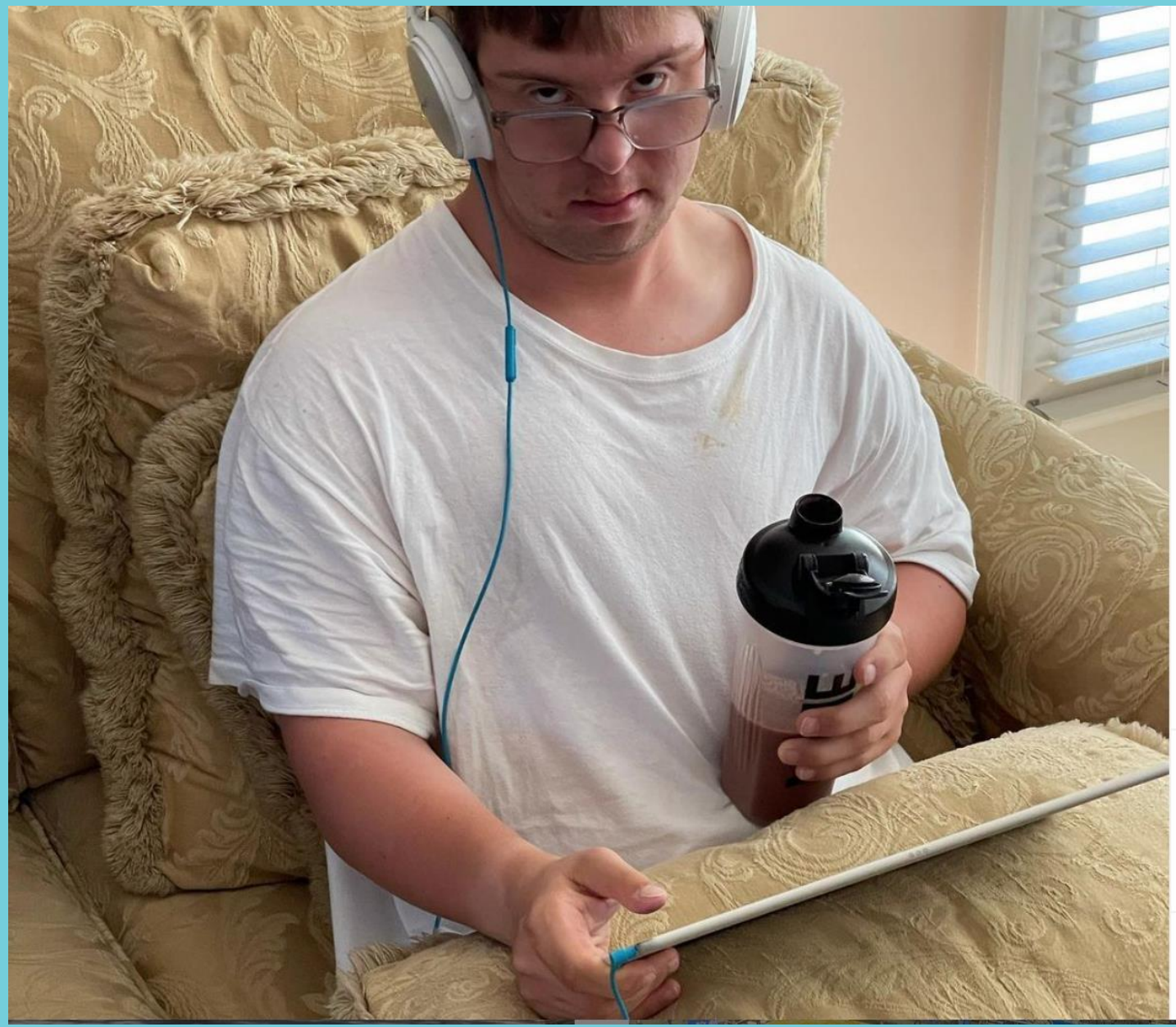
High Rates of Hearing Loss



Many cases of fluctuating and conductive secondary to **otitis media with effusion**
(Nightengale et al., 2017)



- Preschool years: 60-90%
- School-age years: 40-50% (Kong et al. 2017)
- Rates remain high in adulthood



chrisnikic Being sick sucks. Fever, infection, Covid test negative so hoping to get better in time for Ironman 70.3 in Haines City in Sunday. This is day #4 no exercise. This is boring. Miss my training and friends. This is one of the problems with Down Syndrome. Small ear and nose canals make it easy to get an infection but now I get sick less and recover faster. Still plan on doing Haines City Ironman Sunday.

1w

showtimewerner Feel better soon 🙏💪

1w 1 like Reply

View replies (1)

10,098 likes

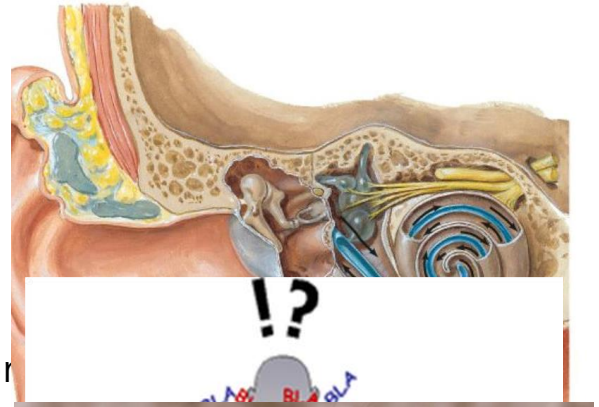
7 DAYS AGO

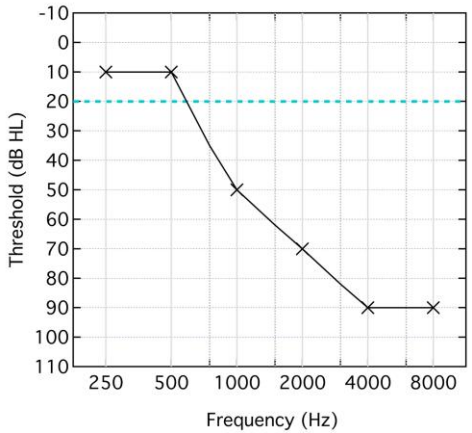
Add a comment... Post

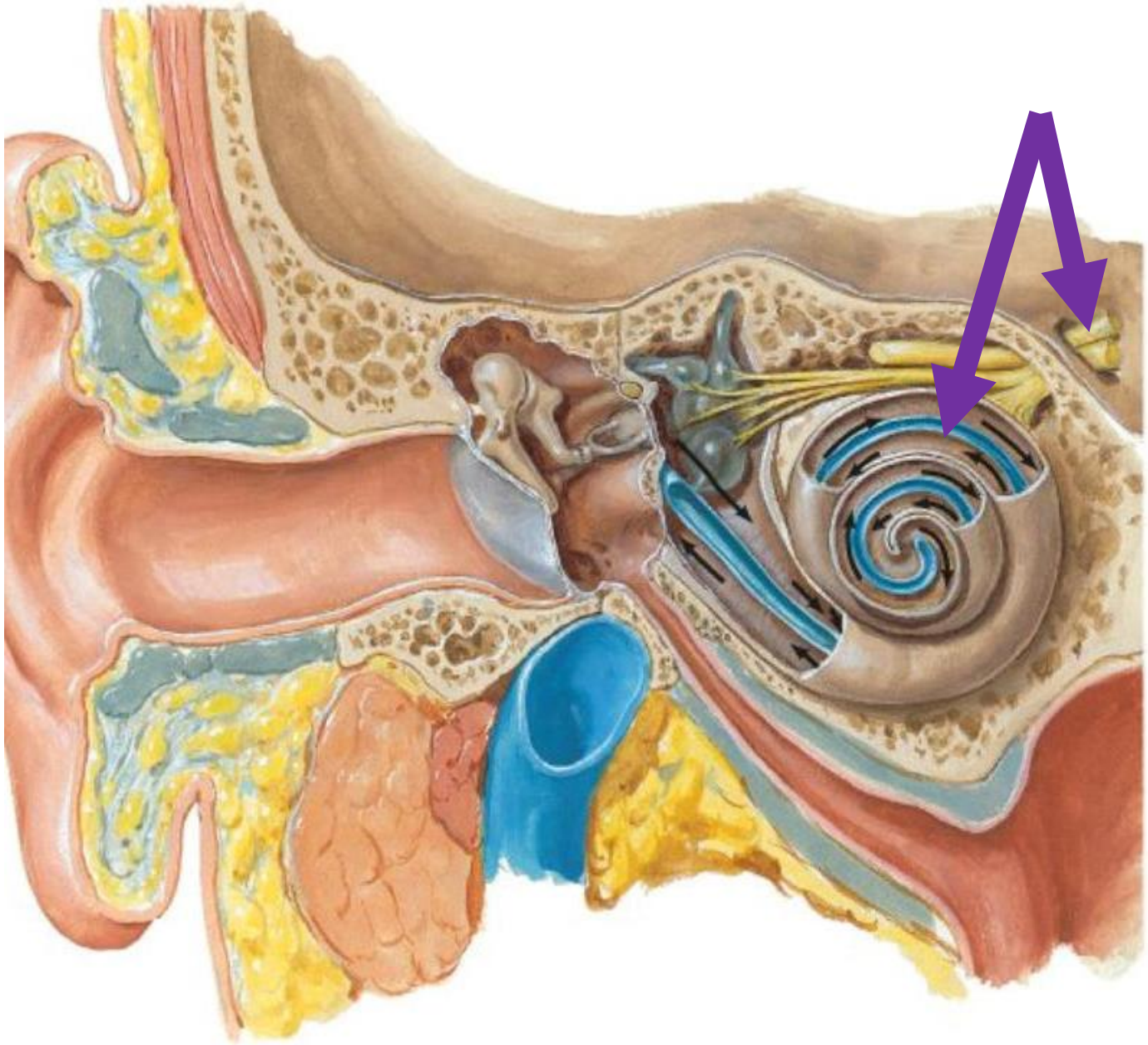
- Small bony structure of inner mouth and nose, but typical muscle mass
- Relaxed muscle function
- Immune system

Many ear infections?

- Acquired ossicular malformation (Bilgin et al., 1996)
- Less language exposure and language delays (e.g., Tor
- Difficulty hearing speech in noise (e.g., Schlider et al. 1994)
- Reduced spatial hearing abilities (e.g., Tomblin & Rance, 2014)
- Persist several years after resolution of the infection
- High frequency hearing loss (Hunter et al., 1996)







Sensorineural Hearing Loss

High Rates of Hearing Loss

High rates of sensorineural hearing loss throughout the lifespan (DeSchrijver et al., 1990)

High Rates of Hearing Loss

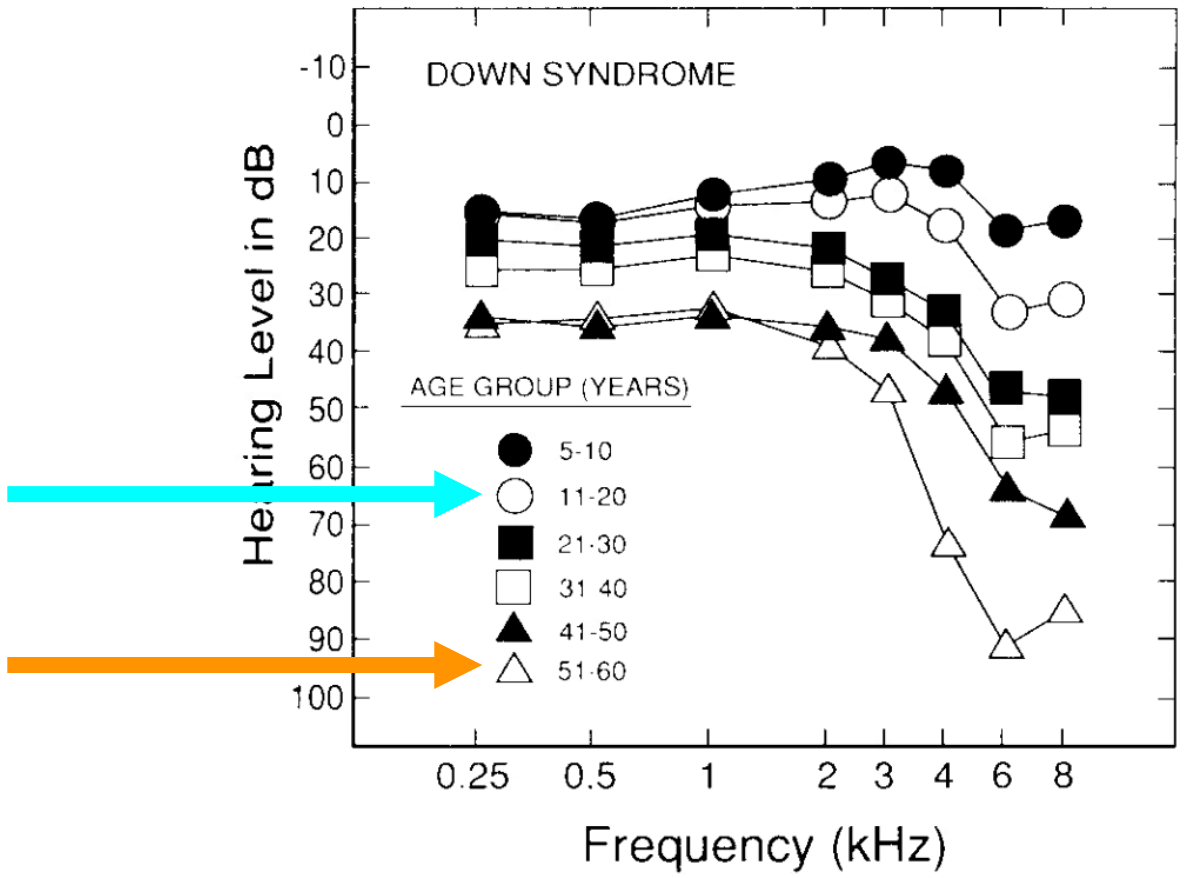
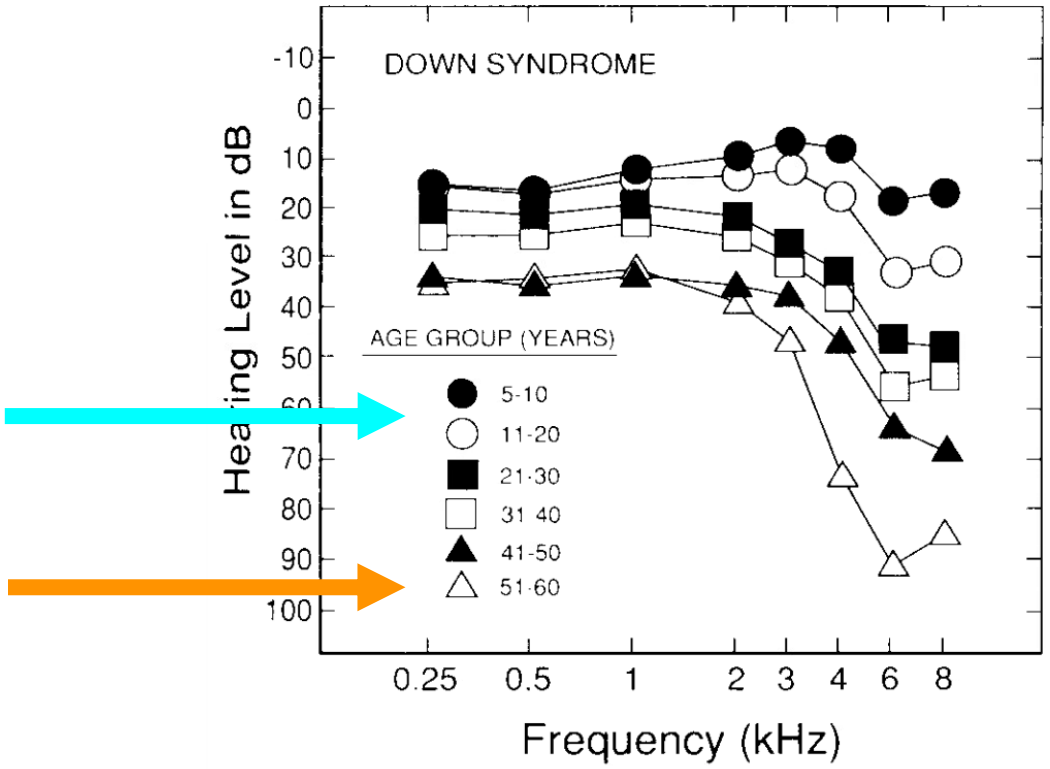


Fig. 1. Mean hearing level as a function of age in Down syndrome.

Buchanan, 1990

Early aging OR result of many ear infections?

Both?



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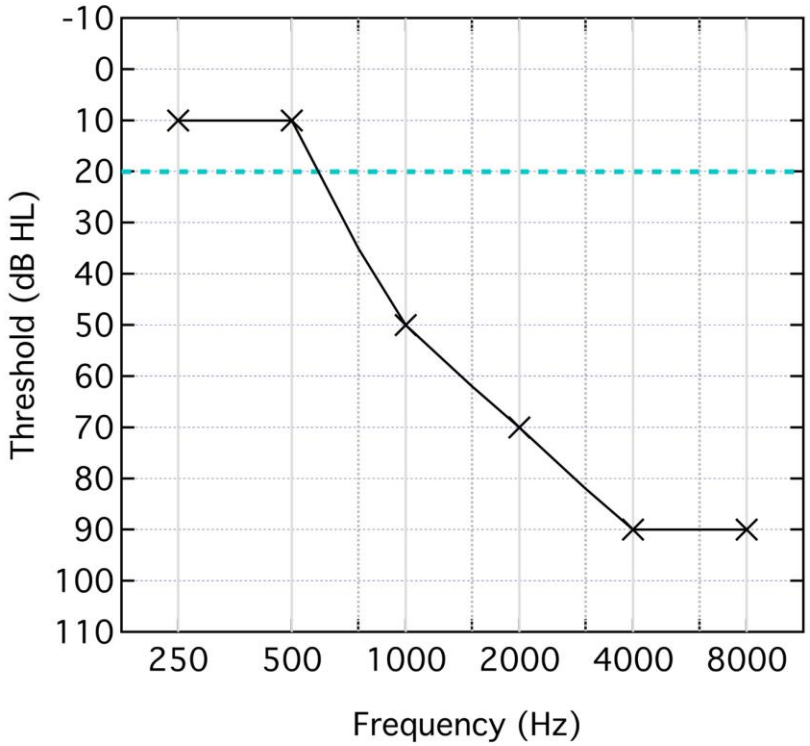


Hunter et al., 1996

Potential Bias

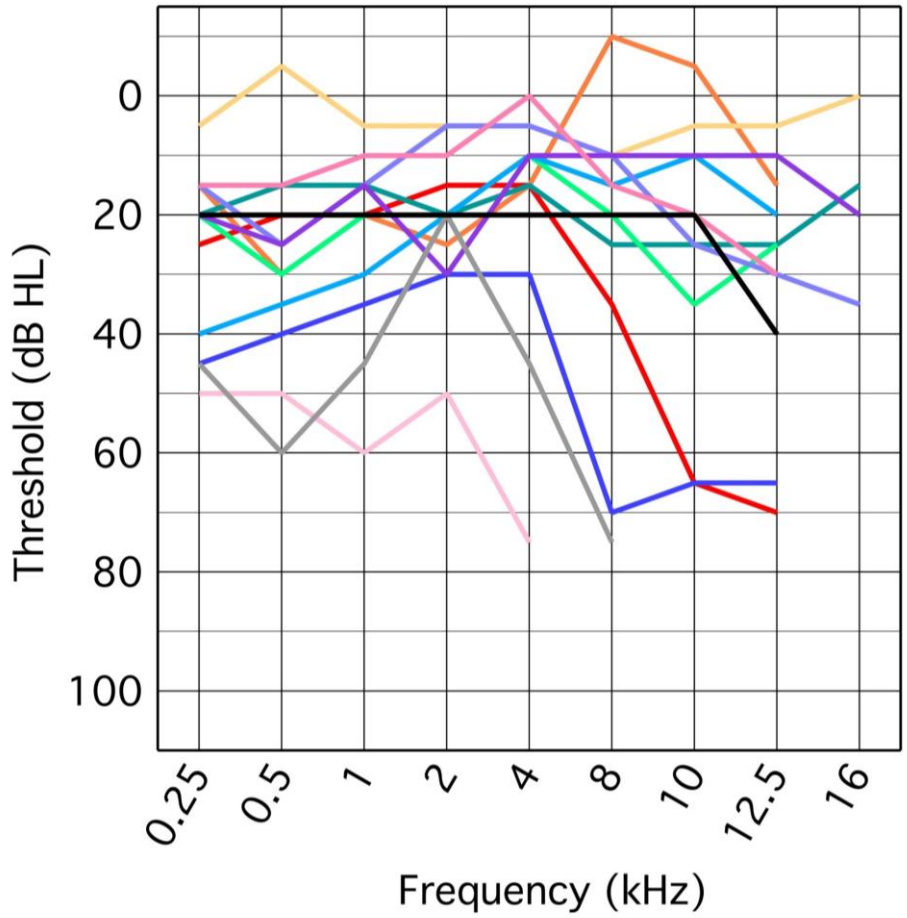
- medical record review
- institutionalized individuals

characterize hearing using observational methods

characterize hearing using observational methods

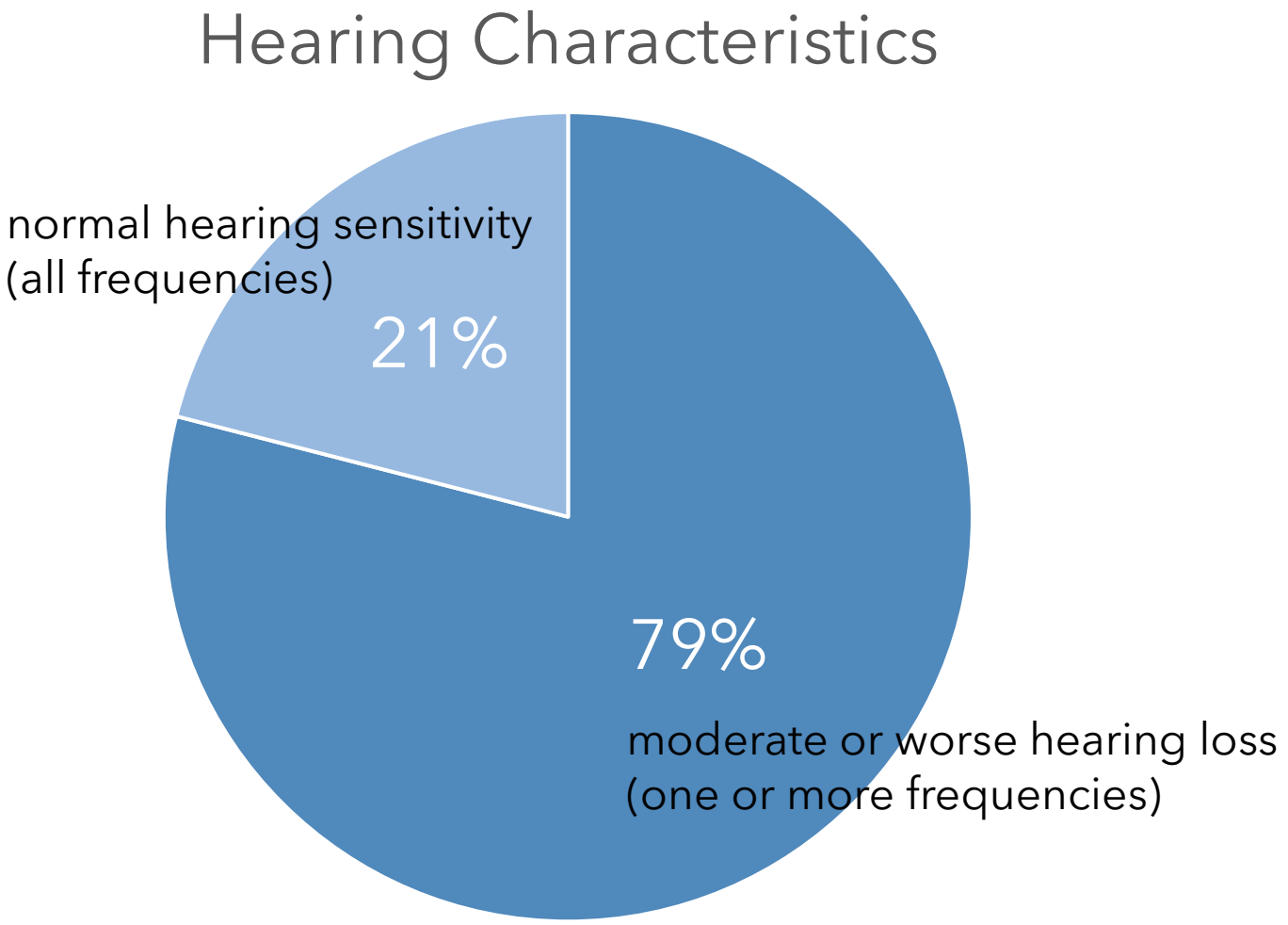
- Participants
  - 18 children with Down syndrome
  - 10 male
- Ages = 5 to 17 years
  - *mean* = 11.0; *stdev* = 4.0

High Rates of Hearing Loss



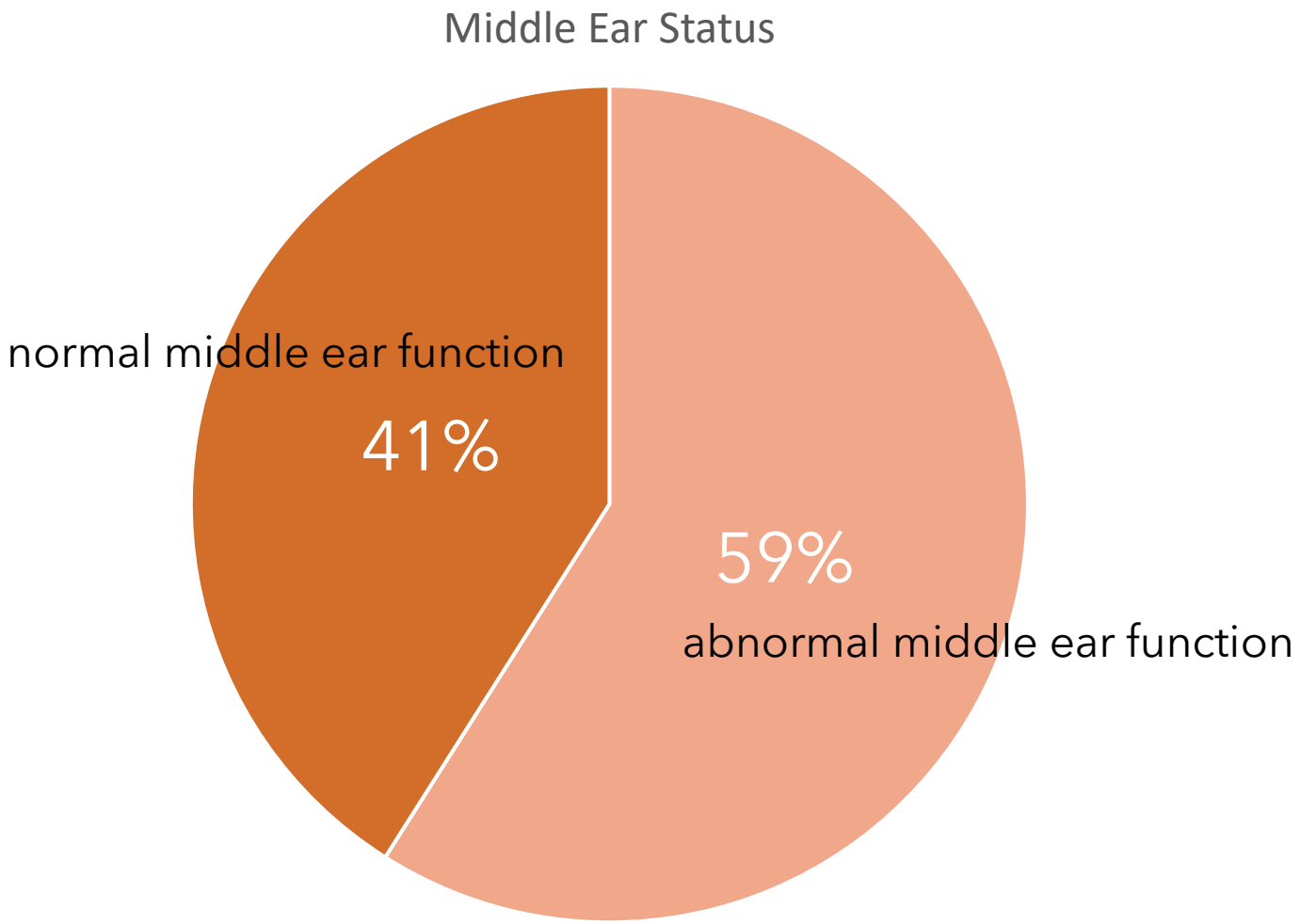
(Porter et al., 2022)

High Rates of Hearing Loss



(Porter et al., 2022)

High Rates of Hearing Loss



(Porter et al., 2022)

Like past studies…

~80% had hearing loss
~60% of participants had middle ear dysfunction

What do think is the worst consequence of untreated hearing loss?

Intervention benefits children with and without additional disabilities.

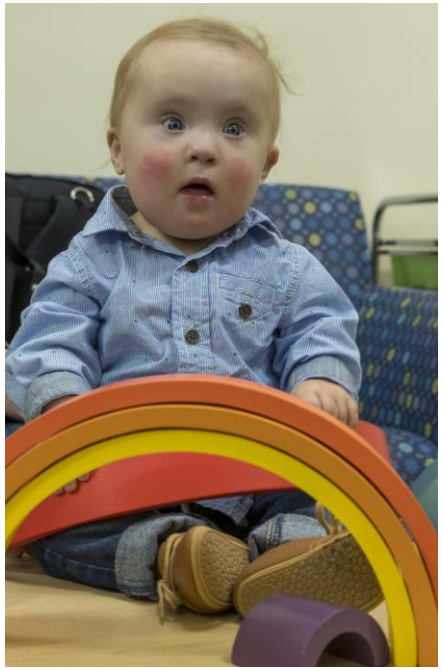
e.g., Cupples et al., 2018; Yoshinaga-Itano et al., 1998

Current recommendations for diagnosis, monitoring for hearing loss, and intervention

Current recommendations for **diagnosis, monitoring for hearing loss**, and intervention

How often should hearing tests be completed?

Hearing Tests: NOT one and done



- Completed every 6 months until hearing sensitivity is established in each ear.
- ABR may be required.
- Once established, behavioral hearing test and tympanograms should be completed annually, or sooner.

e.g., American Academy of Pediatrics, 2022;
Joint Commission on Infant Hearing, 2019

Reflecting on what we have learned.

My child is too little to have a hearing test.

True or False?

My friend with Down syndrome just had a hearing test and everything was normal. They do not need to have another hearing test until they are really old.

True or False?

Current recommendations for diagnosis, monitoring for hearing loss, and **intervention**

Hearing loss: Intervention options

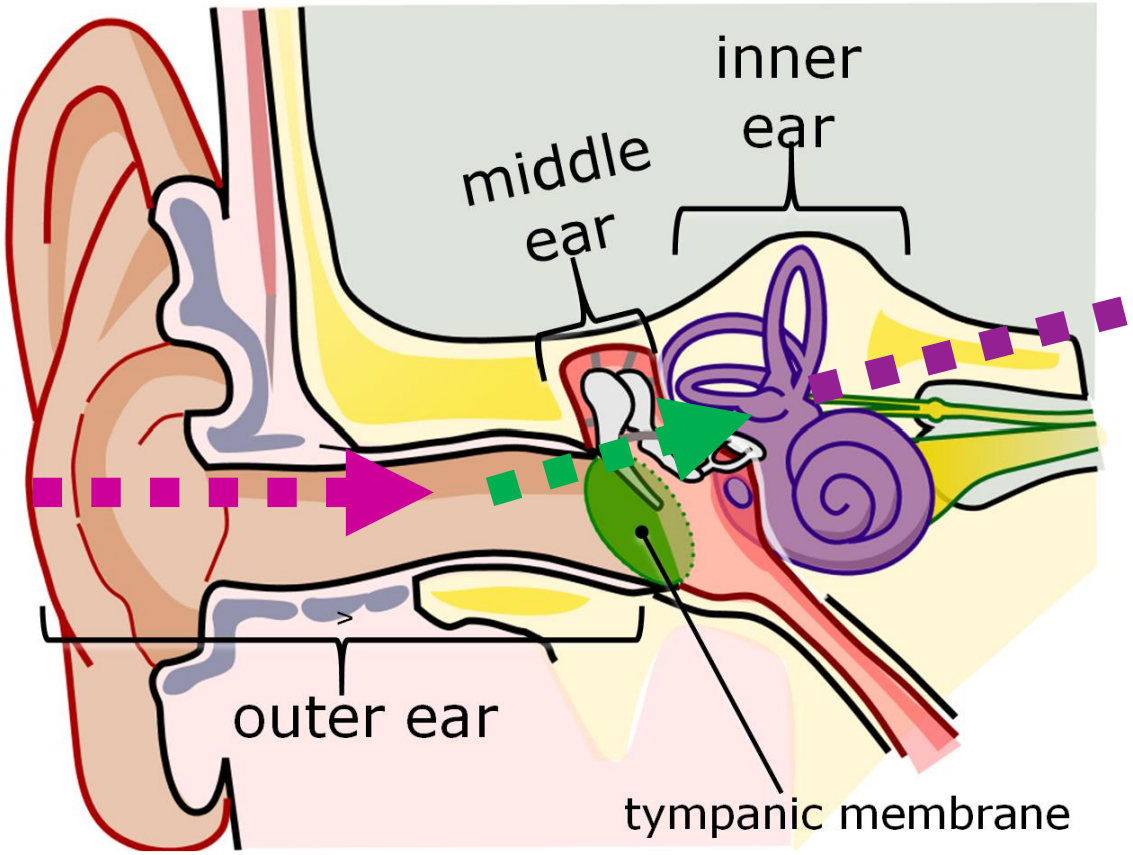
traditional hearing aid
(air conduction hearing aid)

bone conduction hearing aid

cochlear implant

Hearing Aids

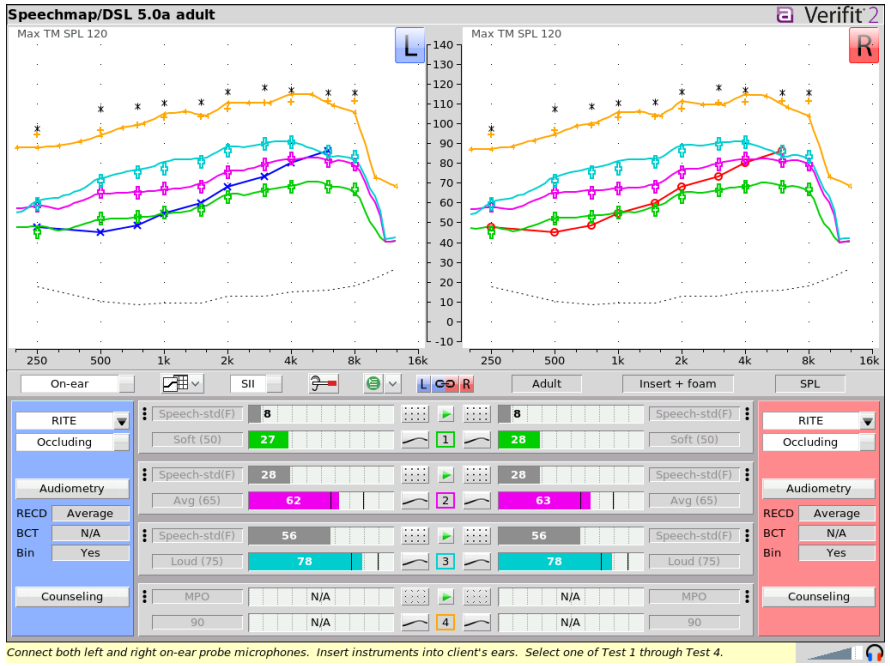
traditional hearing aid



Ears and hearing are very personalized.

Hearing Aid Verification

Essential process to make sure hearing aids are fit according to prescriptive targets.



Every ear is DIFFERENT

Bone conduction device

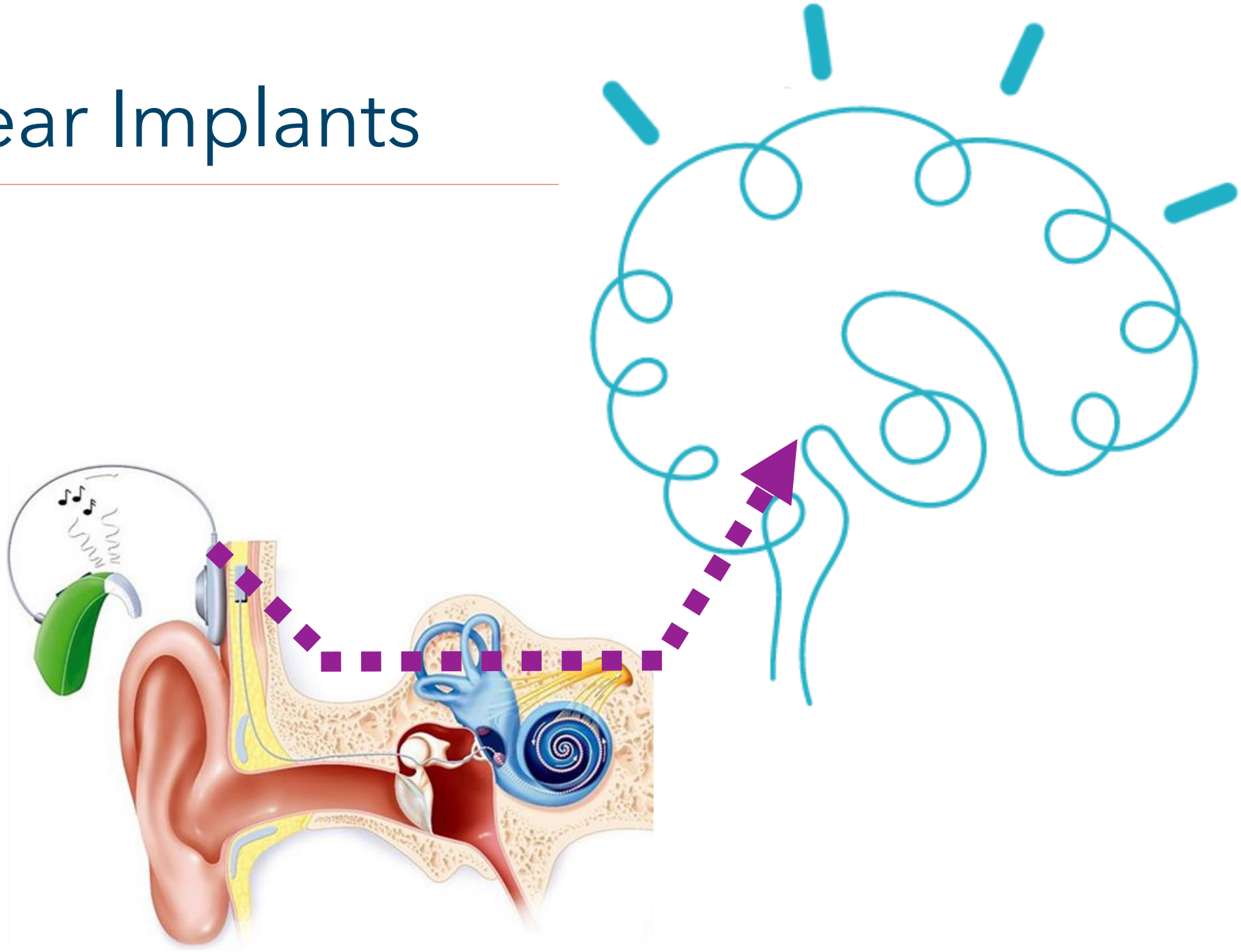
Recommended for children with:

- Structural abnormalities and cannot wear a traditional hearing aid
- Conductive or mixed hearing losses
- Chronic drainage

Cochlear implants

- Considered when a hearing aid is no longer beneficial or powerful enough.

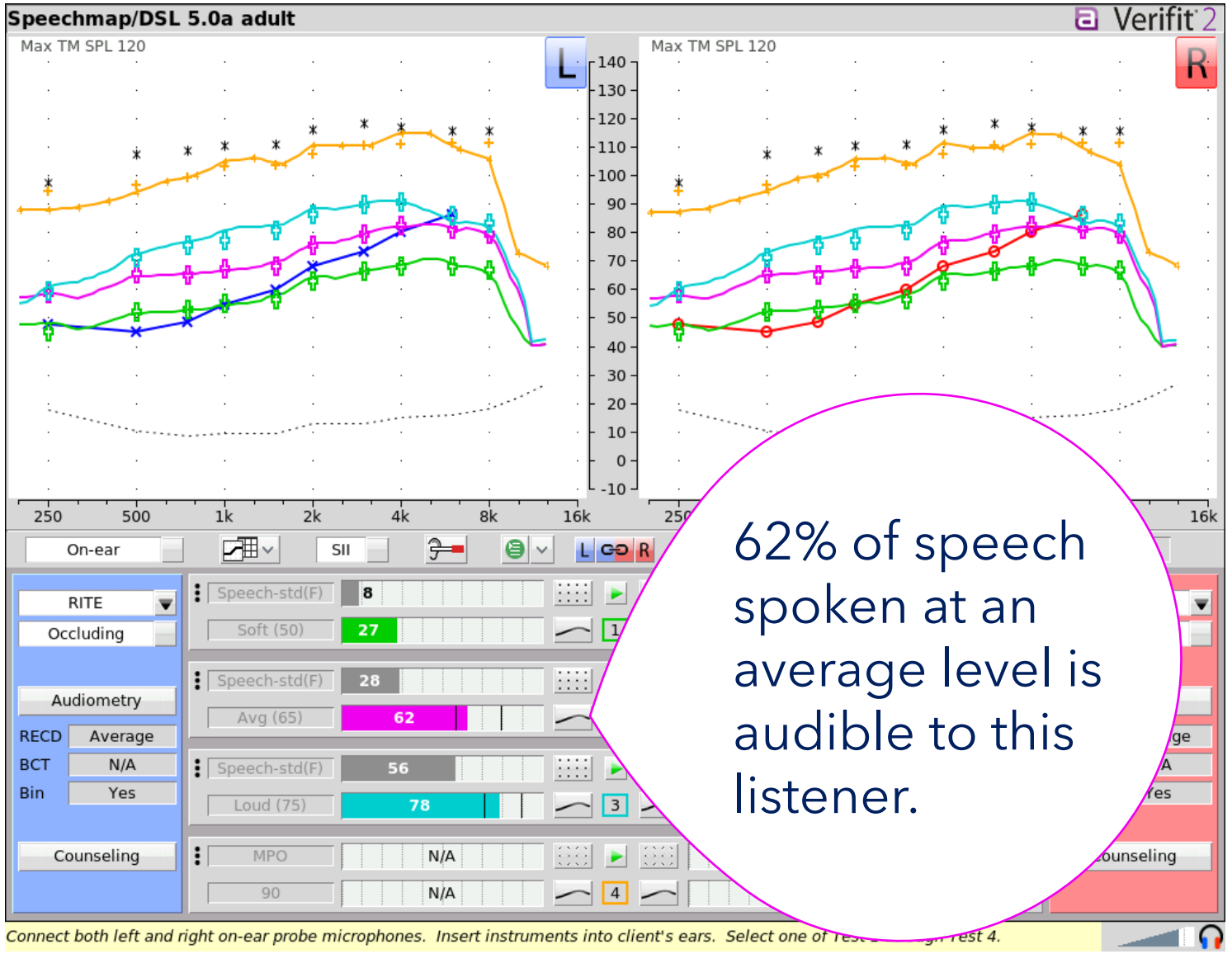
Cochlear Implants



How do I know how when a hearing device is needed?

Speech Intelligibility Index (SII)

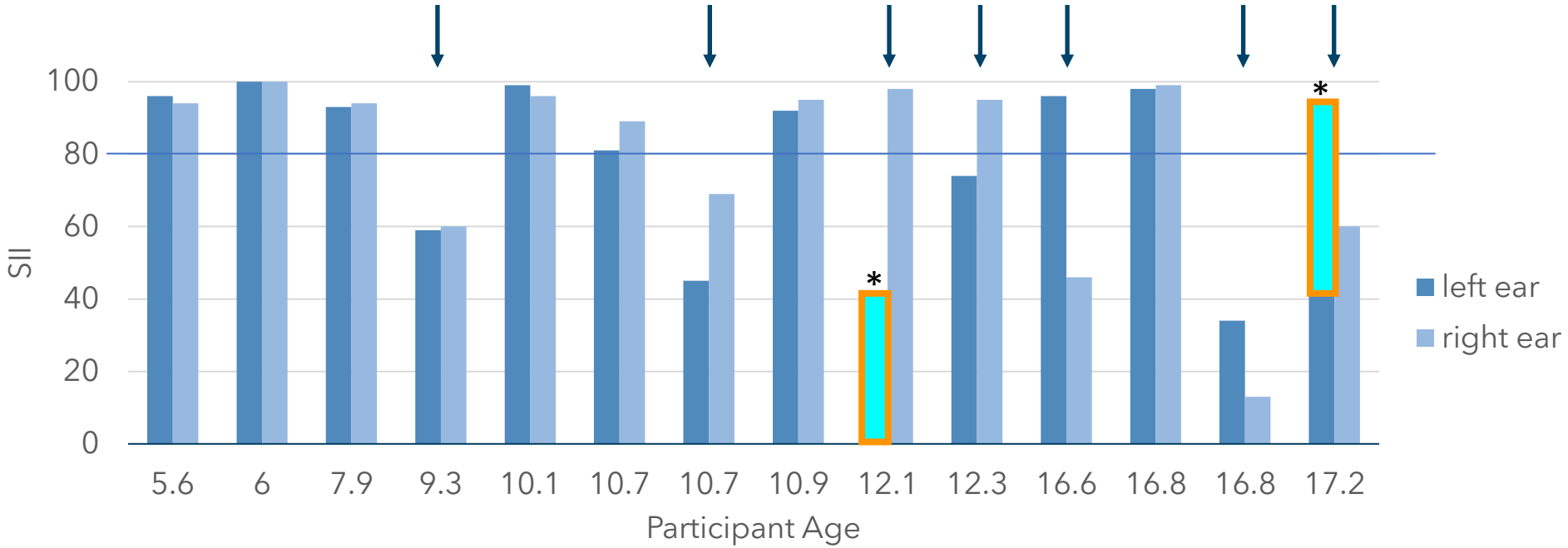
Estimates the percentage of speech a person can hear.



Speech Intelligibility Index (SII)

McCreery et al., 2020

Speech Intelligibility Index (SII)



McCreery et al., 2020

	Left Ear SII	Right Ear SII
average (stdev)	70.8 (32.9)	79.1 (26.0)
median	86.5	94.0
range	0-100	13-100

High Rates of Hearing Loss

7/18 met criteria for hearing aids

2/7 had them

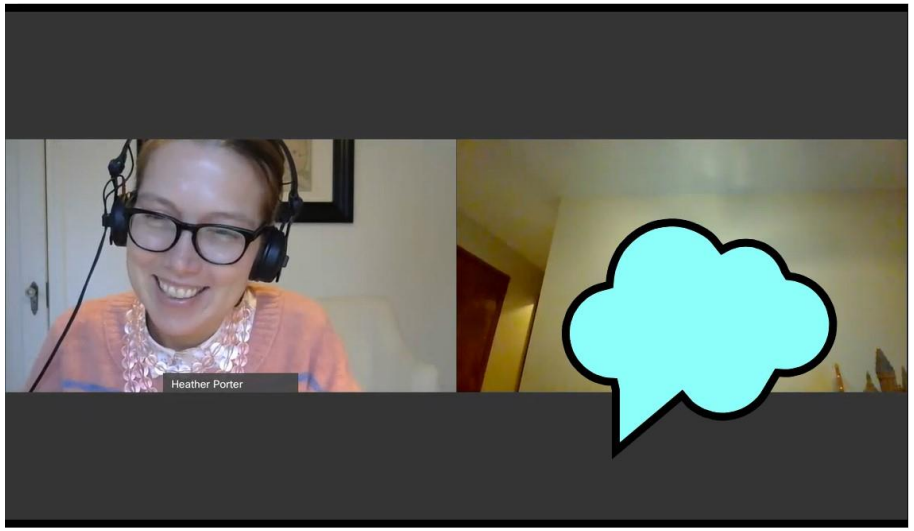
What provides scientific evidence to tell if I need a hearing device?

Common barriers to good hearing

Community Perspectives

- Semi-structured interviews to **investigate listening experiences** and auditory environments of individuals with Down syndrome
- Participants
  - 14 monolingual English-speakers
  - 2 bilingual Spanish/English speakers
- Mother age = 35-67 years
  - mean = 47.8, stdev = 9.1
- Age of child with Down syndrome = 6-40 years
  - mean = 14.0 stdev = 8.2

Community Perspectives



- What are **frequent listening situations** your family member encounters?
- How do you think your family member's **hearing affects their life**?
- What **barriers** to good listening/hearing does your family member experience?
- Reflect on communication moments that were **difficult** for your family member or a potential cause of conflict.

Community Perspectives

PARENT RESPONSES

Our thoughts were accurately represented. We appreciate the work you did.

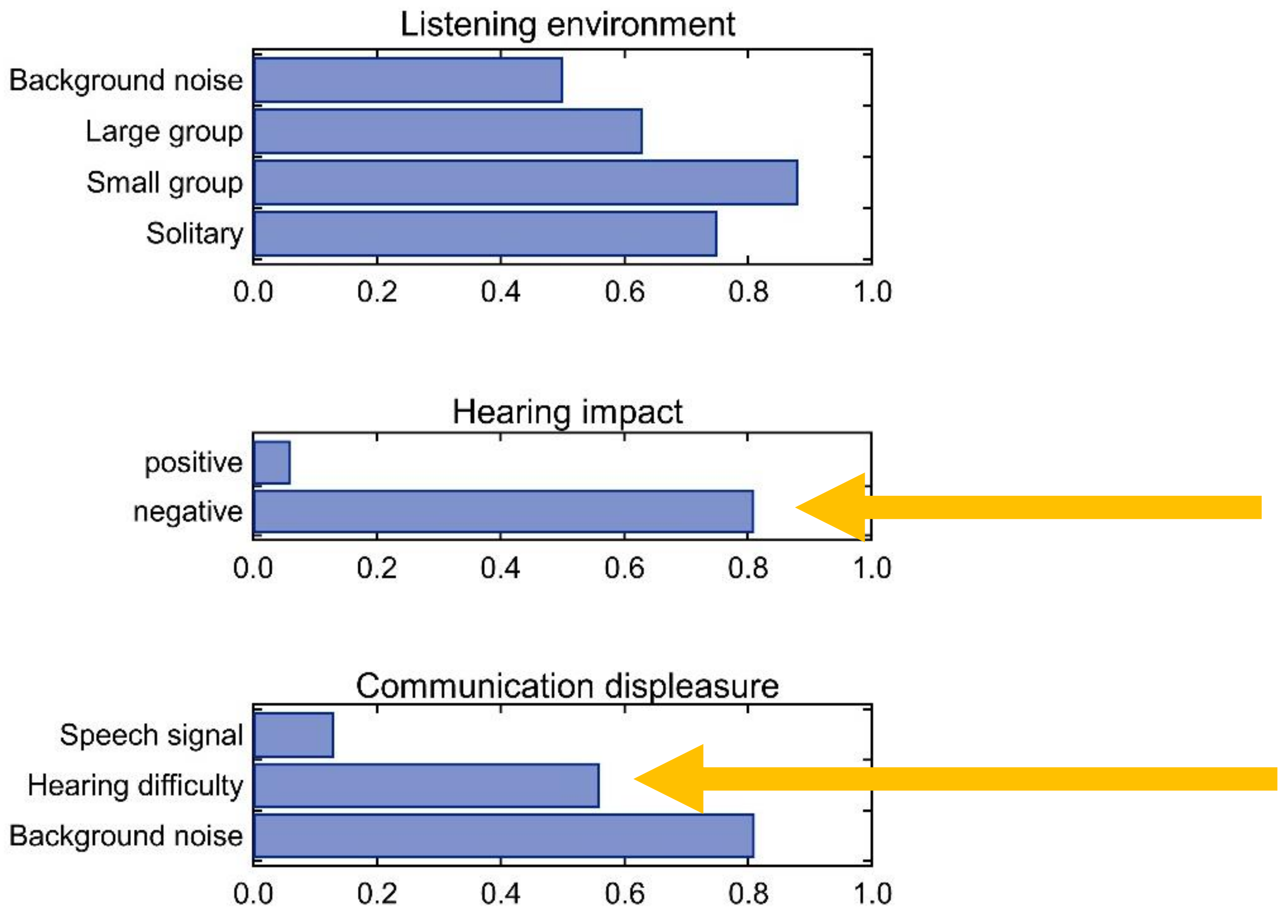
LOVED THE CONCLUSION! GREAT JOB TEAM BOYS TOWN!

I thought the video was beautifully done. I don’t have anything to add.

I feel the information accurately represented my thoughts and I wouldn’t make any change.

I looked over the attachment and watched the video. Everything is well put together! No edits from me. Thanks!


hearing
talking
family
school
people
play
communication
situations
questions
listening
important
loves
help
needs
kids
sounds
understand
thinking
syndrome
person
feel
hard
special
skills
saying
looking
phone
trying
watch
speech
information
music
focus
group
work
year
times


Listening environment
Background noise
Large group
Small group
Solitary
0.0
0.2
0.4
0.6
0.8
1.0
Hearing impact
positive
negative
0.0
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0.8
1.0
Communication displeasure
Speech signal
Hearing difficulty
Background noise
0.0
0.2
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1.0


hearing
talking
family
school
people
play
communication
situations
listening
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help
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important
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person
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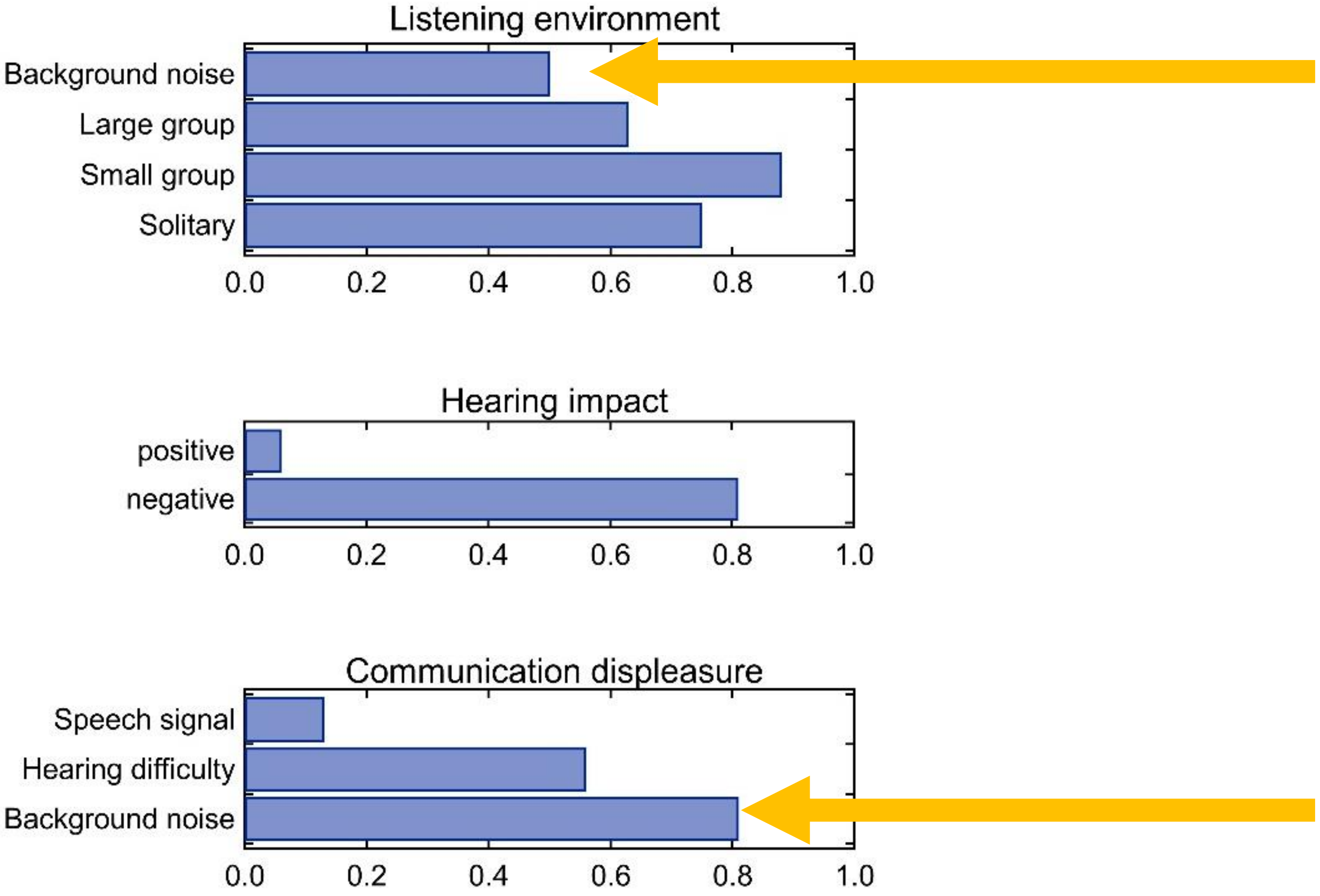
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0.8
1.0
Communication displeasure
Speech signal
Hearing difficulty
Background noise
0.0
0.2
0.4
0.6
0.8
1.0

Communication Tips

For Talking to Individuals with Hearing loss

Face the person directly, and on the same level, whenever possible.

Get the person's attention before you begin talking.

Keep your hands away from your face while talking. If you are eating, or covering your lips, your speech is hard to understand.

Be in the same room. Never talk from another room or while walking away.

Use a clear voice. Speak with a normal tone of voice.
Enunciate.
No shouting!

Reduce background noises when conversing. Turn off the radio or TV.

If a person does not understand what you say, find a different way to say it. Rephrase, rather than repeat.

People often have more difficulty concentrating and understanding if they are ill or tired.

REVIEW

- ✓ Consequences of hearing loss
- ✓ Current recommendations for diagnosis, monitoring for hearing loss, and intervention
- ✓ Common barriers to good hearing

You can advocate for evidence-based hearing healthcare!

Project INCLUDE: study spotlight

Where does the evidence come from?

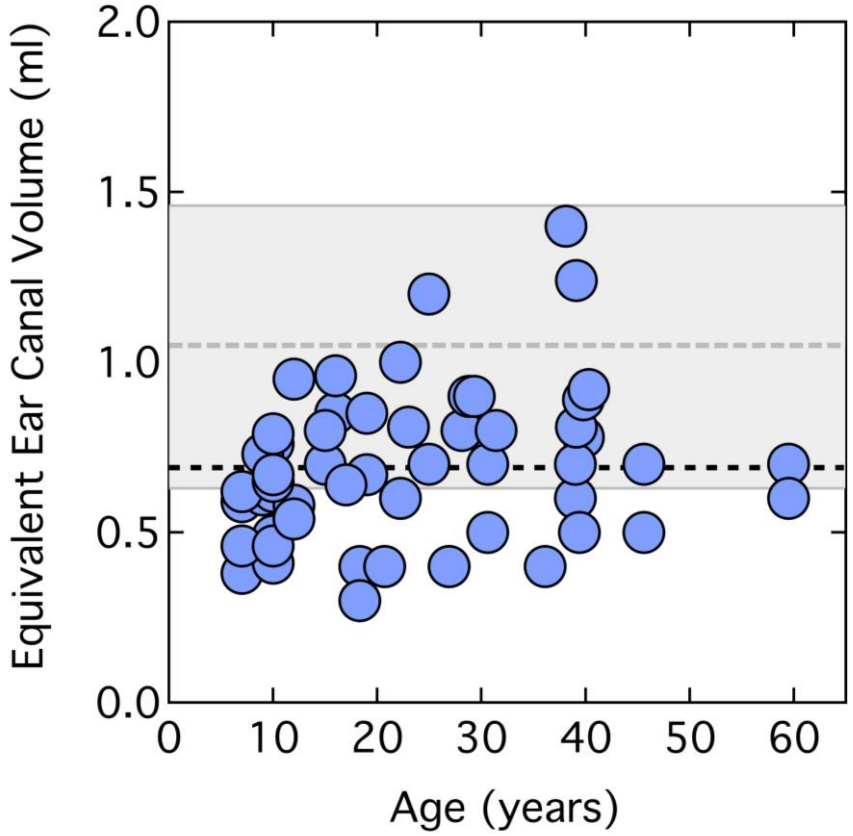
Studies to improve hearing testing

Studies to understand speech perception

Studies to improve hearing testing

Individualized volume measurements

Measuring the impact of individual ear canal sizes during hearing tests



Studies to improve hearing testing

Auditory brainstem response testing

Measuring the impact of individual ear canal size on how the hearing nerve responds to sound

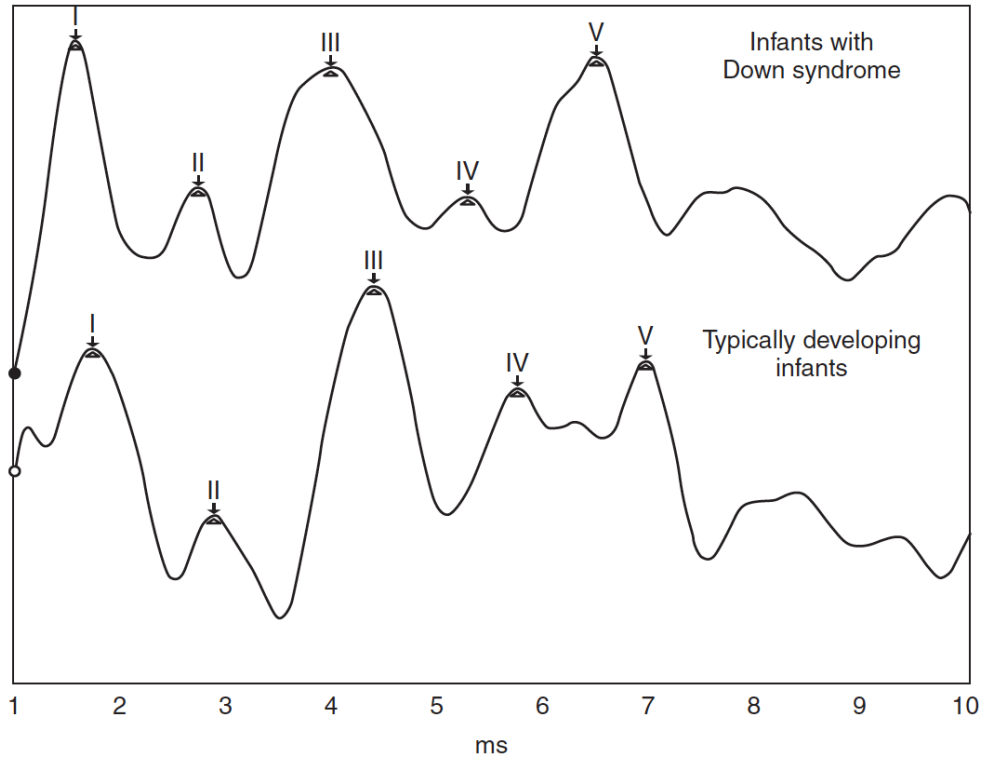


Figure 7.4 Examples of auditory brainstem response (ABR) waveforms to a moderately-high intensity stimuli for healthy infants with Down syndrome and typically developing infants.

Porter & Tharpe, 2010

Studies to improve hearing testing

iPad hearing test

Developing an iPad app for hearing testing that can be used outside of the audiology clinic (e.g., doctors offices, participant homes)

Studies to understand speech perception

Words in noise

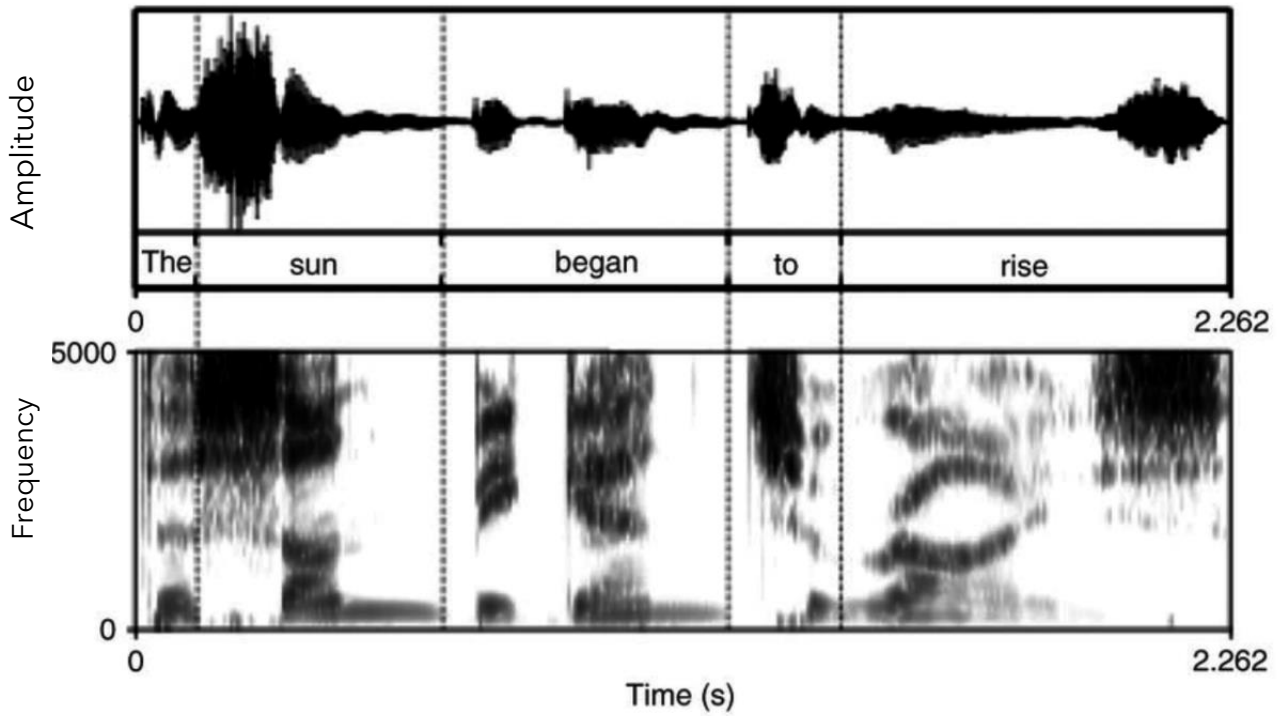
Measuring the ability to hear words that are played at the same time as different background noise

Studies to understand speech perception

Speech sounds

Measuring the ability to detect changes in sounds that are used for speech perception

level changes



pitch changes

Where does the evidence come from?

Volunteers like YOU!

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